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**Current options and recommendations for the treatment of thoracic aortic pathologies involving the aortic arch- an expert consensus document of the European Association for Cardio-Thoracic Surgery (EACTS) and the European Society of Vascular Surgery (ESVS)**

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118	<b>Abbreviations in the text (in order of their occurrence)</b>	
119		
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121	SACP	Selective antegrade cerebral perfusion
122	EACTS	European Association for Cardio-thoracic Surgery
123	ESVS	European Society of Vascular Surgery
124	WC	Writing committee
125	DTA	Descending thoracic aorta
126	IA	Innominate artery
127	LSA	Left subclavian artery
128	TEVAR	Thoracic endovascular aortic repair
129	PAU	Penetrating aortic ulcer
130	AHA	American Heart Association
131	ESC	European Society for Cardiology
132	TAA	Thoracic aortic aneurysm
133	MFS	Marfan syndrome
134	LDS	Loeys Dietz syndrome
135	LCCA	Left common carotid artery
136	TAR	Total arch replacement
137	FET	Frozen elephant trunk
138	IRAD	International Registry of Acute Aortic Dissection
139	IMH	Intramural hematoma
140	CT(A)	Computed tomography (angiography)
141	3D	Three-dimensional
142	CM	Contrast medium
143	ROI	Region of interest
144	HU	Hounsfield units
145	ECG	Electrocardiography
146	MPR	Multiplanar Reformation
147	MIP	Maximum intensity projection
148	VRT	Volume rendering technique
149	CL	Centerline
150	MRI	Magnetic resonance imaging
151	CE	Contrast enhanced
152	MRA	Magnetic resonance angiography
153	GRE	Gradient echo
154	Gd	Gadolinium
155	TOF	Time of flight
156	PC	Phase contrast
157	Gd-BOPTA	Gadobenate dimeglumine
158	NSF	Nephrogenic systemic fibrosis
159	4D	Four-dimensional
160	TWIST	Time-resolved angiography with interleaved stochastic trajectories
161	TREAT	Time-resolved echo-shared angiography
162	US	Ultrasound
163	FoV	Field of view
164	CEUS	Contrast-enhanced ultrasound
165	EVAR	Endovascular aortic repair
166	TEE	Transesophageal echocardiography
167	TTE	Transthoracic echocardiography
168	FOCUS	Focused cardiac ultrasound
169	IVUS	Intravascular ultrasound
170	CPM	Clinical prediction models
171	2D	Two-dimensional
172	RAACP	Right axillary antegrade cerebral perfusion
173	FA	Femoral artery
174	HCA	Hypothermic circulatory arrest
175	CPB	Cardiopulmonary bypass
176	NIRS	Near infra-red spectroscopy
177	Hb	Hemoglobin
178	rSO <sub>2</sub>	Regional cerebral oxygen saturation
179	EEG	Electroencephalography
180	ECI	Electro cerebral inactivity
181	MEP	Motor evoked potentials
182	SSEP	Somatosensory evoked potentials
183	TAA/TAAA	Thoracic/ Thoracoabdominal aortic aneurysm

184	SCI	Spinal cord injury
185	CSF	Cerebrospinal fluid
186	AAA	Abdominal aortic aneurysm
187	CNS	Cerebrospinal fluid
188	UFH	Unfractionated heparin
189	ACT	Activated clotting time
190	POC	Point of care
191	EACTA	European Association of Cardio-thoracic Anesthesia
192	TEG	Thromboelastography
193	ROTEM	Rotational thrombelastometry
194	ET	Elephant trunk
195	TA	Thoracoabdominal
196	CG/PG	Chimney (parallel) graft
197	CABG	Coronary artery bypass grafting
198	LV/RV function	Left ventricular/ right ventricular function
199	MAGIC	Management of Aortic Graft Infection Collaboration
200	TAK	Takayasu's arteritis
201	GCA	Giant cell arteritis
202	STS-PROM	Society of Thoracic Surgeons- predicted risk of mortality
203	STS	Society of Thoracic Surgeons
204	ES	EuroSCORE
205	SIRS	Systemic inflammatory response syndrome
206	PET	Positron emission tomography
207	GC	Glucocorticoid
208	IL	Interleukin
209	TNF	Tumor necrosis factor
210	SVS	Society of Vascular Surgery
211	TAVI	Transcatheter aortic valve implantation
212	SAVR	Surgical aortic valve implantation
213	GERAADA	German Registry for Acute Aortic Dissection Type A
214	MCS	Mechanical circulatory support
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## 1) Introduction

### Purpose

The last decade has substantially broadened treatment options for patients with thoracic aortic pathology involving the aortic arch. Traditionally, treatment of aortic arch pathology was a domain of open cardiac surgery. The advent of combined vascular and endovascular procedures opened a new field thereby enabling treatment in previously operated and in less fit patients. As a subsequent technological leap, branched arch stent-grafts became available and are currently gaining acceptance in the community. Also, open surgery has substantially improved and the increased use of right subclavian artery cannulation and selective antegrade cerebral perfusion (SACP) at warmer lower body circulatory arrest times together with improved monitoring of organ function has substantially contributed to excellent results in these still major operations. Still, neurological complications remain a major concern of all procedures addressing aortic arch pathology irrespective if open or endo. The reduction to a minimum will be one of the major tasks of the future.

Cross linking between cardiac and vascular surgery has amplified knowledge and interestingly enough, although dividing cardiac and vascular surgery into separate units was popular for a time, in many institutions they are ~~being merged~~ **coming together** again to create aortic centers, a trend which should be interpreted as a plea to work together without creating borders between specialties.

Our hope is that in the future, treatment portfolios will be designed by a single group of people working together to understand the natural course of the disease where physicians are doing the right things when it comes to treatment and the entire aortic team follows an anticipative strategy to remain ahead of the disease process.

The purpose of this combined effort of the European Association for Cardio-thoracic Surgery (EACTS) and the European Society for Vascular Surgery (ESVS) was to develop an expert consensus document covering all aspects of aortic arch disease and to provide the community with a pragmatic guide to understand the natural history of the various disease processes, to aid in indicating treatment and to provide support in choosing the right treatment modality in the right patient at the right point in time. Finally, this document aims at harmonizing terminology in acute and chronic proximal thoracic aortic pathology.



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**Classes of recommendation – Table 1**

Classes of Recommendations	Definition
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.
Class II	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.
<i>Class IIa</i>	<i>Weight of evidence/opinion is in favour of usefulness/efficacy.</i>
<i>Class IIb</i>	<i>Usefulness/efficacy is less well established by evidence/opinion.</i>
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.

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The recommendation grade indicates the strength of a recommendation. Definitions of the classes of recommendation are shown in **Table 1**.

Levels of evidence

Level of Evidence A	Data derived from multiple randomized clinical trials or meta-analyses.
Level of Evidence B	Data derived from a single randomized clinical trial or large non-randomized studies.
Level of Evidence C	Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

## Terminology

The WC (Writing Committee) refers to and recommends the use of the definition of attachment zones as provided by “Reporting standards for thoracic endovascular aortic repair” which are also known as “Ishimaru-zones” in the aortic arch **(1) (Figure 1)**.

Regarding anatomical characteristics of the aortic arch, we refer to the classification of type I, type II and type III aortic arch configuration **(2)**. There are three types of aortic arch and they are based on the relationship of the innominate artery to the aortic arch **(3)** Type I aortic arch is characterized by origin of all three great vessels in the same horizontal plane as the outer curvature of the aortic arch. In Type II aortic arch, the innominate artery originates between the horizontal planes of the outer and inner curvatures of the aortic arch. In type III aortic arch, the innominate artery originates below the horizontal plane of the inner curvature of the aortic arch **(Figure 2)**.

Regarding the use of descriptive terms of specific arch configurations such as gothic arch, steep arch angulation and aortic arch radius, no least common denominator could be identified to add a meaningful definition. Therefore, the use of these terms to describe a specific morphology remains subjective.

**Categorization of tears in aortic dissection-** The WC suggests that the terms “multiple entries and reentries” be removed from clinical use and replaced by the wording “most proximal tear”, “communications between lumina” and “most distal tear” in addition to the term “primary entry tear”. This proposed wording should help create a better understanding of the pathophysiology as well as help standardize communication between physicians describing the pathology.

**Phases of acute aortic dissection-** The WC suggests to use the term “acute” for any dissection between the onset of symptoms and 14 days, “subacute” between 15 days and 90 days and “chronic” thereafter.

**Type A, type B and non-A-non-B aortic dissection-** The WC refers to the original proposal from Stanford defining type A aortic dissection as any dissection involving the ascending aorta but refers to type B aortic dissection when only the descending thoracic aorta (DTA) is involved. An arch involvement either by the most proximal tear or by retrograde extension is referred to as non-A-non-B aortic dissection.

**Definition of complications in acute aortic dissection-** The WC uses the wording of the ESVS clinical practice guidelines on the management of descending thoracic aorta diseases defining complicated type B aortic dissection as “the presence of rapid aortic expansion, aortic rupture and/ or hypotension/ shock, visceral, renal or

limb malperfusion, paraplegia/ paraparesis (**spinal malperfusion**), peri-aortic hematoma, Recurrent or refractory pain and refractory hypertension despite adequate medical therapy (4).

The WC also applies this wording for complications in acute type **A** as well as acute non-A-non-B aortic dissection adding pericardial tamponade, acute aortic valve regurgitation, coronary and cerebral malperfusion to the one with either type A or non-A-non-B aortic dissection (5).

**Aortic arch replacement in various extent-** When referring to aortic arch treatment, qualitative and semi-quantitative statements should be avoided. Given the rising number of patients receiving open and endovascular therapy, it seems reasonable to refer to the treatment based classification using the terminology "zones 0-4" when describing surgery on the aortic arch. Again, "distal arch aneurysm" covers a wide range of anatomical variations and replacing the arch using a FET with an anastomosis proximal to the left carotid artery and selective re-implantation using separate grafts is not adequately covered in the current definitions.

One notable exception is the term "hemi-arch" which has been widely used for decades even if it also covers a wide range of surgical strategies from just replacing the ascending aorta and performing an open distal anastomosis to resecting the entire concavity of the arch down to the proximal DTA.

For the purpose of this paper, total arch replacement is defined as replacing the entire aortic arch- or excluding it from circulation as it is the case when using the frozen elephant trunk (FET) technique- from the offspring of the innominate artery (IA) to a point beyond the offspring of the left subclavian artery (LSA). Reimplantation or revascularization of the supra-aortic branches can be performed in many ways and the method used is not part of the definition of total arch replacement. To facilitate communication and to harmonize the standards of reporting, defining total arch replacement as replacing (or excluding from circulation) aortic zones 0 to 2 (or beyond) seems reasonable. All other procedures on the arch should be named partial arch replacement.

**Residual dissection after type A repair-** The chronic dissected state of aortic segments distal to the proximal repair is defined as "residual dissection after type A repair".

**Chimneys, snorkels, periscopes-** The WC refers to chimneys, snorkels and periscopes using the term "parallel grafts".

## Organization

**Aortic team definition**—The WC advocates that an aortic team—should be closely involved from diagnosis to treatment and finally follow-up being led by cardiac and vascular surgery in collaboration with anesthesiology, cardiology, radiology and genetics. A major advantage of surgery as the leading specialty is that surgeons do have the knowledge of linking radiographic findings to tissue quality which is a major component when opting for open or endovascular treatment.

Additionally, centralization of care of aortic arch pathologies in large centers is recommended because this is the only way to effectively understand the natural course of the disease, provide the entire range of treatment options under one umbrella and treat potential complications of each individual therapy (6). A streamlined emergent care pathway (24/7 availability without diversion), adequate transportation and transfer capabilities as well as rapid activation of the multidisciplinary team must be available.

There is growing evidence, that there is clear correlation between numbers and outcome also in aortic medicine (7–12). With regard to imaging, it is clear that the ability to obtain a hybrid-room setting is limited in many hospitals. However, few trade-offs should be made as adequate intraoperative imaging forms the basis of reliable delivery of quality.

Finally, a structured surveillance of all patients either before they reach the criteria for treatment or after treatment is strongly emphasized. One reason is quality control, another one is the potential to develop aortic pathology in non-treated upstream or downstream aortic segments.

**RECOMMENDATION 1** Decision making for the treatment of aortic arch pathologies by an aortic team is recommended. Class I Level C

**RECOMMENDATION 2** Centralization of care for aortic arch pathologies is recommended. Class I Level C

**RECOMMENDATION 3** Treatment of elective aortic arch pathology is recommended to be performed in specialized centers providing open and endovascular cardiac and vascular surgery on site only. Class I Level C

**RECOMMENDATION 4** Continuing follow-up of patients with aortic arch pathologies before and after treatment in a dedicated outpatient clinic is recommended. Class I Level C

372 RECOMMENDATION 5 A hybrid-room with a fixed imaging system is recommended for thoracic endovascular  
373 aortic repair involving the aortic arch. **Class I Level C**

## 2) Natural course of the disease and underlying pathologies

The vast majority of aortic arch pathologies are based on either aneurysm formation or dissection. While dissection on the basis of previous aneurysm formation is rare, it is the main driver for accelerated growth during follow-up. Isolated aneurysm of the aortic arch is rare and most arch aneurysms that ultimately lead to surgical intervention are caused by aneurysms or dissections of either the ascending or the DTA which at some point extend into the arch or by penetrating aortic ulcers (PAU).

### Natural course of the disease

Population based studies have shown that 60% of thoracic aortic aneurysms (TAA) occur in the root or the ascending aorta, 40% in the DTA and 10% include the aortic arch with some extending into more than one thoracic aortic segment (13). There is no controlled trial that specifically looked at the natural history of aortic arch disease. Several papers discussing the fate of the aortic arch do so by almost exclusively citing data that was derived from either observations on the ascending or the DTA. Moreover, contemporary observational studies and registries are heavily biased by the fact that many patients with aneurysm diameters exceeding the threshold for surgery recommended by the current guidelines do in fact undergo surgery (14). Therefore, there is a tendency towards facing dissection in patients with smaller diameters that had not yet reached the threshold for surgery. Conversely, some patients present with large aneurysms that by far exceed the current recommendations for surgery but have not yet dissected. Most papers dealing with aortic diameters and risk for dissection base their conclusions on post-dissection diameters. Due to the formation of intra- and peri-aortic hematoma, measuring the post-dissection diameter is not reliable. A study looking at patients with acute type A dissection that for some reason previously underwent imaging of the aorta has shown that aortic diameter increases by about 30% at the time of dissection (15). This clearly indicates that diameter at the time of presentation itself is not the sole predictor of the risk of dissection.

The 2010 American Heart Association (AHA) (2) and 2014 European Society for Cardiology (ESC) (16) guidelines refer to various publications that focused on interventions in arch aneurysms or dissections, especially regarding hybrid procedures but the 2014 ESC guidelines do not cite a single paper on the natural history of arch aneurysm and the 2010 AHA guidelines refer only to the 1997 paper from the Yale cohort (17). Data from the Yale aortic database has demonstrated an average annual growth rate of 1mm for ascending aortic aneurysms and 2.9mm for descending aortic aneurysms. Nevertheless, growth rates vary according to the underlying disease and the

absolute size of the aneurysm. Larger aneurysms tend to grow faster. It is important to realize that 95% of patients with TAA are asymptomatic until the first event. Calculating the risk for dissection or rupture is difficult but a large study including 721 patients with TAA demonstrated an annual risk for dissection or rupture of 6.9% in patients with an aneurysm diameter greater than 60mm. Five-year survival in patients with TAA not undergoing intervention was only 54% (18,19).

There are only few reports that focus specifically on the aortic arch. In a small study including 45 patients over a 14-year period with a mean follow-up of 37 months, average annual growth rate was 2.5mm per year but varied widely between 0 and 16mm. During the study period, 22% of patients suffered from rupture. The authors calculated that aneurysms with an annual growth rate of >5.5 mm per year have a 67% likelihood of rupture compared with 8.3% in patients with a growth rate of <5.5 mm per year. Furthermore, in their study, aneurysm size >6.5 cm and hyperlipidemia correlated with more rapid expansion. In a multivariate analysis, growth rate was the sole independent risk factor for aneurysm rupture (OR 1.43; 95% confidence interval, 1.06-1.92; p=0.018) (20). Despite the current evidence is low, there seems to be no justification to conduct a prospective randomized trial comparing natural history to treatment.

It has been shown that 21% of patients with TAA have a relative with an already known aneurysm and that patients with familial occurrence of TAA grow faster than those with sporadic forms (2.1mm per years vs. 1.6mm per year; ascending and DTA combined) (21). This is an important aspect of thoracic aortic disease and rapid progress is currently made in identifying genetic mutations causing TAA. Over the past decade, the medical community has slowly accepted the idea that patients presenting with aortic aneurysm and/or dissection are part of a wide spectrum of genetically mediated diseases that present in syndromic as well as non-syndromic forms. Marfan syndrome (MFS) has long been the only seriously considered differential diagnosis in terms of a heritable disorder of connective tissue in patients with aortic aneurysm. It has been shown that aneurysm formation in MFS is driven by excessive transforming growth factor- $\beta$  (TGF- $\beta$ ), a ubiquitous cytokine in most mammalian cells and involved in cellular proliferation and differentiation. Loeys and Dietz identified a subset of patients sharing certain features such as a bifid uvula, hypertelorism and marked tortuosity of the vessels that had not been typically associated with MFS. The group identified mutations in the gene encoding for the TGF- $\beta$  receptors 1 and 2 as the causative mutation (22,23). Identifying Loeys-Dietz syndrome (LDS) as a separate entity was important as patients with LDS suffered from acute aortic dissection at aortic diameters that had not been considered a cut-off to proceed to surgery in MFS patients. Meanwhile several different mutations in patients within the spectrum of LDS have been identified. Preliminary data suggests significant differences in the risk of acute dissection in these patients. Data from the Johns Hopkins group showed that a significant number of LDS patients had to undergo



interventions on the aortic arch after elective root replacement, something that has been rarely seen in Marfan patients.

With the advent of high-throughput sequencing techniques, more and more causative mutations in non-syndromic forms of type A aortic dissection have been identified. It has been shown that 11% to 19% of patients without (known) genetic defect have first degree relative with type A aortic dissection. Identifying the causative mutation in patients presenting with type A aortic dissection has a direct impact on the indication for surgery, the extent of surgery, and the prognosis of the patient, as well as his relatives.

#### **Underlying pathologies- Aortic arch dissection**

According to the Stanford classification of aortic dissection, a dissection is considered to be a type A dissection if the ascending aorta is involved, regardless of the location of the primary entry tear. According to this definition, a dissection in the aortic arch is generally considered a type B dissection. But as 90% of the type B dissections occur distal to the LSA, the majority of data on type B dissection does not apply to aortic arch dissection (4). Nevertheless, the notion of "non-A-non-B" dissections needs to be established (Figure 3).

**Some studies have implicated anatomical variants as predisposing factors for dissections with entries in the aortic arch. In a study including 157 patients (24) that underwent surgery for acute type A aortic dissection, 14% of patients had either a common origin of the IA and the left common carotid artery (LCCA) or an origin of the LCCA from the IA and the rate of arch entries in this group was significantly higher compared to patients without this pattern (59% vs. 13%,  $p<0.001$ ). Furthermore, the presence of this arch pattern was associated with a higher rate of post-operative neurological injury (OR 4.9; 95% CI, 1.635-14.734;  $p=0.005$ ).**

#### **Type A aortic dissection**

The fate of the aortic arch in patients with type A aortic dissection is strongly correlated with the extent of the initial surgery. It has been clearly shown that not replacing the entire ascending aorta results in a high rate of re-operations. Therefore, performing at least a primary entry tear-oriented hemi-arch replacement is recommended.

The additional burden of replacing the entire aortic arch as an adjunct to elective or emergent proximal repair is not very well defined and makes comparison with patients undergoing secondary total arch replacement difficult. Most papers reporting on outcomes after surgery for type A dissection or those dealing with re-intervention after proximal repair do not discuss arch related morbidity and mortality separately (25,26). The major risk factor for the

need of re-intervention on the aortic arch and distal aorta after repaired type A dissection is a patent false lumen. Furthermore, pseudoaneurysm or dehiscence at the level of the distal anastomosis has been described as a frequent cause for re-operation. Therefore, several groups began to advocate total arch replacement and implantation of a frozen elephant trunk in addition to proximal repair in type A dissection.

Interestingly, Asian groups tend to favour a more aggressive approach and mostly recommend total arch replacement during initial surgery for type A dissection. It has been discussed whether this is also due to a more favorable anatomy in the Asian population and a more pronounced atherosclerotic burden in western countries which increases the risk for stroke during total arch replacement. In 2009, a Japanese group published one of the very few reports comparing hemi-arch replacement with an open distal anastomosis to total arch replacement (TAR) with implantation of a frozen elephant trunk (FET) (27). In 120 patients presenting with acute type A dissection, mortality was only 4% with no new cerebral events and a survival of 95% at 5 years in the FET group compared to 69% in the hemi-arch group.

A Chinese-American collaboration focusing specifically on patients with type A dissection and an entry tear in the arch analyzed 104 patients who underwent FET and total arch replacement and compared them with 728 patients undergoing surgery for type A dissection with entry tears elsewhere. Operative mortality was 8.6% with a 2.9% paraplegia rate. Stroke rate was surprisingly low with 1.9%. In this series, survival and freedom from late adverse events was 89% and 85% at 8 years, respectively, after a mean follow-up of  $5.6 \pm 2.6$  years. Compared to other series, the time from onset of symptoms to surgery was quite long with  $4.7 \pm 3.5$  days. Furthermore, computed tomography (CT) results after a mean of  $4.6 \pm 2.9$  years postoperatively were only available in 65 patients, but showed complete false lumen obliteration in 63 patients. The authors concluded that type A dissection with entry in the arch can be treated safely by FET and total arch replacement and provides durable results (28). Unfortunately, a true comparison with patients undergoing less extensive surgery was not performed.

Data from patients with MFS have shown that the extent of arch surgery during the initial intervention did not influence the need for thoraco-abdominal repair during follow-up. These data suggest that it is the dissection itself that drives the need for re-operations in these patients and that the aortic arch is only one of many segments that have to be repaired over the years (29). In a large series of MFS patients, it was shown that there was no significant difference regarding the rate of re-operation in patients with persisting dissection in the DTA after TAR compared to those without (30). The rate for re-interventions was 50% in both groups at 10 years. Nevertheless, the rate of re-operation was higher in patients with a dissection in the aortic arch where only the ascending aorta was replaced compared to those patients without a dissected arch. Therefore, in the rare cases where the dissection is confined to the aortic arch, complete exclusion of the dissection may reduce the need for re-

interventions and should be attempted. The principal importance of closing the primary entry tear during the index procedure and the differences in the natural history of the disease if the primary entry tear has been effectively closed or not have been previously described (31)

## **Type B dissection**

The International Registry of Acute Aortic Dissection (IRAD) investigators compared patients with and without retrograde extension of type B dissection (32). Retrograde extension into the aortic arch occurred in 16.5% of patients. There were no differences in the rate of patients presenting with complicated type B dissection. In this registry, there were no differences regarding choice of treatment by the participating centers. Patients with and without arch involvement received best medical treatment only in 53.7% vs. 56.5% ( $p=0.68$ ), endovascular treatment in 32.8% vs. 31.1% ( $p=0.78$ ), open operation in 11.9% vs. 9.5% ( $p=0.54$ ), or hybrid approach in 1.5% vs. 3.0% ( $p=0.70$ ), respectively. Furthermore, there was no difference in in-hospital mortality in patients with (10.7%) or without (10.4%) retrograde arch extension ( $p=0.96$ ). Five-year survival was similar with 78.3% and 77.8%, respectively ( $p=0.27$ ). Unfortunately, this study did not look at those patients that not only had an arch involvement but also had their primary entry tear in the arch.

A few years ago, it was proposed that patients with an entry at the inner curvature of the distal aortic arch have a higher risk to suffer from complicated type B dissection compared to those with an entry on the outer curvature (33,34). At that time it was speculated that the LSA may represent a natural barrier for progress of the dissection into the aortic arch. In this series, the incidence of primary complicated type B aortic dissection was 3 times higher in patients with an entry in the lesser curvature compared to those with an entry in the outer curvature (61% vs. 21%,  $p = 0.003$ ). Interestingly, a Japanese study with a total of 224 patients with type B dissection found that in multivariate analysis an entry at the outer curvature of the distal aortic arch was associated with a higher need of late open aortic surgery, aortic interventions and aortic events after a mean follow-up of  $6.0\pm 4.1$  years (35). However, it has to be stated, that there are several clinical scenarios where the location of the primary entry tear remains either unclear or a matter of discussion, e.g. in combination with IMH. This might be owed to the quality of imaging or simply to a masked disease process. Serial adequate imaging may unmask the exact location of the primary entry tear within the first days after the acute event as can TEE help in elucidating the exact location (36).

## **Non-A-non-B aortic dissection – type B dissection involving aortic arch**

Both Stanford and DeBakey classifications do not address the clinical scenario when the aortic arch is dissected, but not the ascending aorta (37). In the ESC 2014 aortic guidelines the comments on Stanford classification regarding arch dissection in patients with non-dissected ascending aorta is missing (16). The 2010 AHA guidelines recommends to categorize patients with descending aortic dissection and entry within the arch as proximal type B dissection (2). Distal type B dissection refers to descending aortic dissection and entry distal to the LSA (2). The evolution of the term Non-A-non-B aortic dissection can be more seen as a kind of evolution of understanding of the pathophysiological process having been initially described in 1994 (38). In a recent study including 43 patients with descending aortic dissection and dissection components in the aortic arch, authors found 21 patients with entry in the DTA and 22 patients with entry within the aortic arch (39). The incidence of non-A non-B dissection was 11% among all patients with acute aortic dissection. Non-A non-B dissection patients presented with a common origin of the IA and LCCA in 28% and an arch origin left vertebral artery in 16%. The overwhelming majority of patients underwent aortic repair. Emergency aortic repair due to malperfusion or aortic rupture was necessary in 29% descending-entry and 36% arch-entry type patients. Another 43% descending-entry and 36% arch-entry patients required aortic repair within 2 weeks after dissection onset due to rapid aortic growth, aortic rupture, new organ malperfusion or persisting pain. All patients, except for 1, required repair for aneurysm at follow-up. Overall in-hospital mortality in acute non-A non-B dissection patients was 9%. The highest in-hospital mortality of 37% was observed in arch-entry patients who underwent emergency surgery.

Clinical presentation, treatment and outcome in non-A non-B dissection patients are different from those commonly reported for patients with acute type B dissection. The involvement of arch in dissection process of the DTA seems to have an important impact on clinical course and outcome, therefore it is reasonable not to categorize these patients as type B, but as non-A non-B aortic dissection.

## **Aortic Intramural Hematoma**

The ESC guidelines define aortic intramural hematoma (IMH) as a circular or crescent-shaped thickening >5mm of the aortic wall with absence of a dissecting membrane, intimal disruption or false lumen flow (16). **The ESVS guidelines define intramural hematoma as the presence of blood within the aortic wall without intimal disruption or an identifiable entry point on imaging (4).** While current guidelines see IMH as a separate entity, distinguishing between IMH and dissection may not always be possible in clinical practice. There is certainly a time-dependent variable with regard to diagnosis as patients frequently present with new intimal lesions 24 to 48

hours after the initial imaging studies were performed. The current definition of IMH may be challenged as more sophisticated imaging methods will be able to identify more primary entry tears and therefore identify more IMH as a precursor of acute aortic dissection.

Some of the predictive factors for disease progression that have been proposed for patients with IMH without associated ulcer or intimal erosion include involvement of the ascending aorta, aortic diameter >50 mm in initial imaging as well as persistent pain. Predictors of disease progression in patients with IMH and an associated aortic ulcer or intimal erosion include increase of associated pleural effusion, recurrent pain, ulcer located in the ascending aorta or arch with initial maximum ulcer diameter >20mm or more and initial maximum ulcer depth >10mm (40,41). In a German multicenter study, 60% of IMH patients revealed evidence of significant progression and 20% developed overt dissection within 30 days of hospital admission (42).

Data are particularly scarce on IMH in the aortic arch. In a 2012 publication on IMH from the IRAD investigators, the authors analyzed 178 patients. 42% of whom presented with type A and 58% with type B IMH. In 24 (13%) of these the most proximal extent was in the aortic arch. Separate analysis of these patients showed that 16 were medically managed, 4 underwent surgery, 2 received endovascular treatment, and 2 had hybrid interventions. There were 3 deaths (12.5%) in the population and the authors concluded that this group had a slightly higher mortality and increased need for interventions than patients presenting with type B IMH (43).

### **Penetrating Aortic Ulcer**

The current ESC guidelines on aortic disease define PAU as an ulceration of an aortic atherosclerotic plaque penetrating through the internal elastic lamina into the media. It is thought that PAU represents 2 to 7% of all patients with acute aortic syndromes. While there are no controlled studies regarding the natural history of PAU in different settings, reports have shown that PAU can result in the development of true aortic aneurysm, IMH or aortic dissection. Patients presenting with PAU frequently have a high atherosclerotic burden. Risk factors for PAU include advanced age, male gender, tobacco smoking, hypertension, coronary artery disease, chronic obstructive pulmonary disease, and presence of abdominal aneurysm. In a study from the Mayo Clinic including 105 patients, ulcerations were located in the DTA in 94 patients, in 11 patients in the aortic arch and 10% presented with PAUs in multiple locations. Interestingly, the rate of PAUs located in the arch was significantly higher in the group of patients that was asymptomatic compared to those that were symptomatic (20% vs. 5%,  $p=0.03$ ) (44).

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614 This data is in line with a large series of 388 patients from the Philadelphia group presenting with PAU where  
615 6.8% of patients had PAUs located in the aortic arch. The authors report a higher number of open repairs in this  
616 patient group but there is no data regarding specific outcome parameters **(45)**. Indications for intervention  
617 according to the current guidelines include persistent or recurrent pain, contained rupture, rapid growth, periaortic  
618 hematoma and pleural effusion. It is thought that in asymptomatic PAU a diameter >20mm and a neck >10mm  
619 have a higher risk of progression and early intervention should be evaluated.

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622 **Recommendation for open and endovascular interventions based on aortic diameter**

623

624 Given the paucity of data on the natural history as well as the varying results of open surgery, there are few  
625 recommendations regarding the optimal timing of surgery solely based on the diameter of the arch. The current  
626 guidelines recommend surgery in isolated arch aneurysms at a diameter of 55mm. Both, the AHA and the ESC  
627 guidelines acknowledge the fact that the indication for surgery in arch aneurysm is strongly influenced by the  
628 overall vascular situation and especially the diameter of the adjacent ascending and descending aortic segments.  
629 In the majority of patients, this will determine the threshold for intervention.

630

631 **RECOMMENDATION 6 Treatment of isolated aortic arch aneurysms should be considered at a diameter of**  
632 **55mm. Class IIA Level B (2)**

### 3) Imaging and diagnostic work-up

#### Computed Tomography Angiography

CT is the most commonly used imaging modality to assess the aorta and has many advantages over other imaging modalities. Currently, it remains the modality of first choice (46). It is able to quickly acquire high spatial resolution three-dimensional (3D) images of the aorta and surrounding structures and enable diagnosis and aid in planning treatment.

The acquisition should start cranially to the aortic arch and include the supra-aortic branches- ideally the circle of Willis, and extend caudally to the level of the femoral heads. A scan prior to contrast administration ("native") is performed in some institutions for some questions, e.g. to rule out IMH. A total of 50 to 120 ml of contrast medium (CM) is generally needed (0.5 to 0.7 g of iodine per kilogram of body weight) (47,48). At the CT console a region of interest (ROI) marker is placed in the thoracic aorta. When the contrast enhancement reaches a certain density threshold (e.g. 120 Hounsfield Units, HU) within the chosen ROI the start of the scan is delayed for a few seconds (depending on the scanners' speed) to perform data acquisition at the correct position in the ideal moment of the arterial phase. If needed for evaluation of e.g. organ perfusion, a second scan in the venous phase may be acquired after a delay of 60- 90 seconds upon arrival of contrast.

CT data can be acquired with reference to the electrocardiogram (ECG) signal to provide images of each phase of the cardiac cycle, in order to minimize the artifacts from cardiac pulsation and aortic wall motion, which requires a low pitch down to 0.2, i.e. a slow-moving table. There are two techniques to obtain an ECG- gated CTA, pro- or retrospectively (49). Artifacts from an incompressible patient, bowel or breathing motion are not compensated. To describe cardiac or vessel motion during an R-R'-interval a maximum of twenty 3D data sets of the entire cardiac cycle can be gained using *retrospective* triggering. This allows reconstruction of max. 20 3D CTA. This dynamic CTA provides information on aortic movement and dynamic changes in aortic perfusion. However, radiation dose of retrospectively triggered or gated CTA is much higher in comparison to conventional CTA (49,50). The use of dual source technology and the high pitch that can be achieved with this technique (up to 3.4), may overcome the need for ECG-triggering and thus reduce radiation dose, without loss of diagnostic accuracy (51).

Post-processing of axial CT data is possible using multiplanar reformation (MPR), maximal intensity projection (MIP) and volume rendering technique (VRT) (49,52). MPR allows for generation of an arbitrarily angled cross-section within the entire three-dimensional data set. Such MPRs allow a better visualization and appreciation of anatomical and pathological structures (49,53). Semi- or full automatic centerline (CL) analysis are used to

improve length measurement accuracy and to achieve diameter perpendicular to the CL (53). Aortic diameter measurements must always be obtained using MPR reconstruction on planes perpendicular to the aortic flow direction ("double-oblique" technique) (54).

## **Magnetic Resonance Imaging**

Magnetic resonance imaging (MRI) can provide 3D images of the aorta and surrounding structures with high contrast enhancement and high spatial resolution. MRI has obvious advantages over CT including superior soft tissue contrast, the absence of ionizing radiation, and the ability to depict and quantify functional parameters. Combining anatomical and functional information in a single acquisition means that MRI can potentially provide a more comprehensive evaluation of thoracic aortic disease. The relatively long acquisition times however limit its use in the acute setting.

Magnetic resonance angiography (MRA) is the most commonly used MRI technique for both pre- and post-procedural imaging of the thoracic aorta. CE (Contrast enhanced) MRA techniques rely on the T1 shortening effect of Gadolinium(Gd)-chelate contrast agents in blood to generate high intravascular signal, instead of exploiting the inherent motion of blood flow as in the flow-based time-of-flight (TOF) and phase-contrast (PC) techniques. Thanks to this different approach the vascular signal generated with CE-MRA is not hampered by the numerous flow-related artifacts that can degrade the flow-based MRA techniques (55,56). One of the more effective compounds for vascular contrast is Gd-BOPTA (Gadobenate dimeglumine), which has been proven to perform better than the standard compounds due to weak binding to serum albumin (57). Some issues were raised regarding the occurrence of a syndrome named nephrogenic systemic fibrosis (NSF), that limits the applicability of CE-MRA in patients with renal insufficiency (58). There is active research going on investigating the relevance of Gd-deposition in the human body after contrast enhanced exams, especially in the brain (59,60). Today, no clinical symptoms have been described associated with intracerebral Gd-deposition.

The use of phased array coils provides the additional benefit of markedly shortening image acquisition times or, with the use of parallel imaging schemes, of acquiring higher spatial resolution image sets in the same time period (61,62). As with CTA, the vascular enhancement is a transient and dynamic process, hence the critical element to be set for a CE-MRA is the proper timing for the image acquisition.

Dynamic MRA provides temporal information during the heart cycle that can be visualized as a dynamic display, thereby adding a fourth dimension, 4D CE-MRA. Its acquisition is typically combined with a Gd-based CM injection while a sequence of 3D volumes is acquired over time including fat suppression over time (49,63,64).



Fast GRE (Gradient echo)-sequence covers the entire aorta allowing high temporal resolution of e.g. 2-4s/volume and an interpolated spatial resolution of 1mm<sup>3</sup> at a static magnetic field strength of 3 Tesla. This fastest, time-resolved MRA techniques are available, with two common acronyms for this approach: TWIST (time-resolved angiography with interleaved stochastic trajectories) and TREAT (time-resolved echo-shared angiographic technique) (65).

Cranial magnetic resonance imaging can be used besides intracranial Doppler ultrasound (US) to assess Circle of Willis completeness, which helps predicting risk of insufficient cross-flow and stroke. Time-resolved MRA of the thoracic aorta is the optimal method to study mobility, stiffness and dynamics of dissection membranes, as well as resulting static or dynamic large vessel occlusion mechanisms. Similar to intracardiac flow dynamics in valvular disease, true and false lumen antegrade, retrograde and turbulent flows should be imaged using MRA as 'gold standard'.

## **Ultrasound**

US techniques have a low small-field-of-view (FoV) compared with CT and MRI. Ultrasound is also constrained by not being able to image through bone or gasses/ air but US can provide functional information with high temporal resolution. Contrast enhanced US (CEUS) is currently being performed using microbubbles as i.v. exogenous CM, e.g. for endoleak detection during endovascular aortic repair (EVAR) follow-up (66). Both trans-esophageal (TTE) and trans-thoracic echocardiography (TTE) can be performed bedside with a low incidence of complications. Using a variety of imaging projections, the aorta and its major branches can be visualized. More recently 3D techniques have been developed that can provide further information regarding the aorta and valve function although its clinical incremental value has not yet been fully assessed.

**Ultrasound can add important dynamic and functional insights in to the disease process at several levels also with regard to aortic branches of first order such as the supraaortic, visceral, renal and iliac/femoral vessels.**

Intravascular ultrasound (IVUS) provides dynamic information regarding both true and false lumen and allows to detect false lumen thrombosis with higher sensitivity and specificity than TEE. Because of its invasiveness, the use of IVUS is limited to intraoperative guidance.

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736 **Diagnostic Work-up in Aortic Arch Disease – Emergency Repair Setting**

737 The diagnostic workup in preparation of emergency aortic arch repair, in most cases acute Stanford Type A or  
738 non-A-non-B dissections, also focuses on selecting the most effective, most durable, and safest operative and  
739 perfusion strategy, however with less time available and a limited diagnostic workup of supraaortic and  
740 intracranial collateral flow. CTA, TEE in the operation theatre, and sometimes supraaortic Duplex US of carotid  
741 arteries are possible providing sufficient information to be able to plan and to treat.

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744 **RECOMMENDATION 7** Preoperative assessment of aortic arch pathologies with Computed Tomography  
745 Angiography is recommended as first line imaging modality Class I Level C

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747 **RECOMMENDATION 8** Assessment of patency and morphology of the circle of Willis is recommended where  
748 treatment involves the aortic arch Class I Level C

749

750 **RECOMMENDATION 9** Assessment of the extracranial supraaortic vessels down to the level of the femoral artery  
751 bifurcation is recommended where treatment involves the aortic arch Class I Level C

752

#### 4) Risk stratification, patient and treatment approach selection

Risk constellations and case mix in patients with aortic disease are no less heterogeneous than in the cardiac surgical population. Currently available modalities for perioperative risk assessment like STS Risk Model for Mortality (STS-PROM) (67) or ES I and II (68) have been well validated for cardiac surgery, but not for aortic disease and its surgical and endovascular treatment options. Thus, STS-PROM and ES are inappropriate risk prediction tools for patients with aortic arch pathologies and procedures. The same holds true for other, unmodeled severity scores.

Since clinical prediction models (CPM) are indispensable for any risk stratification in patients undergoing invasive procedures, their lack for aortic arch pathologies hampers comparison of prospective study results, database analyses, therapies and of institutional and health care systems performance. In this field, development of a dedicated CPM and risk score remains therefore an unmet need. In recognition of the increasing frequency and complexity of thoracic aortic medicine, the STS has recently formed a task force on aortic surgery and has added aortic pathology as a module in order to collect data for CPM development and further research (69).

Patient selection and selection of the treatment approach-Aortic arch pathology in various extent without any further affection of the cardiovascular system is the exception and not the rule. Despite that several underlying pathologies leading to the final common path of aneurysmal formation/ lesion development, the algorithm to diagnose concomitant cardiac and vascular conditions should be standardized in all patients being evaluated for treatment and finally, outcome of this diagnostic algorithm should also have an impact on the final treatment strategy.

Each patient should undergo TTE or in case of remaining need, TEE. Coronary angiography is recommended in all patients in need for open surgery whereas non-invasive testing might be regarded as sufficient in selected cases scheduled for endotherapy in the absence of symptoms indicative for coronary artery disease. In candidates for endovascular treatment with a past medical history of coronary artery disease, additional diagnostics should be considered to quantify the severity of the underlying concomitant condition.

**Supraaortic branches should be evaluated by supraaortic US and there is definitive need for evaluating cerebral cross-flow and the patency of the Circle of Willis.** Finally, a CTA should evaluate the entire aorta including her branches of first order. Harmonization of the aforementioned diagnostic should then lead to a recommendation for treatment be it open surgery, combined vascular and endovascular procedures, a full

endovascular approach or to a recommendation for conservative treatment in case that the remaining risk of concomitant conditions outweigh the potential benefit of treatment.

#### **5) Monitoring during aortic arch repair**

As for any major cardiovascular surgery, standard monitoring includes non-invasive and invasive hemodynamic, respiratory, anesthesia, temperature, coagulation and laboratory monitoring. Additional monitoring techniques for aortic arch procedures should be selected according to specific requirements of patient, surgeon and interventionist, in order to help preserve hemodynamics and organ function, and to support procedural management (2).

#### **Transoesophageal Echocardiography**

TEE offers real-time 2D and 3D morphological and functional cardiovascular assessment as a semi-invasive imaging modality. Echocardiography systems used in aortic arch programs should include options and probes for epiaortic and epicardial US, for Doppler and 2D interrogation of supraaortic and peripheral vessels, as well as for US-guided vascular access. There is consensus in current guidelines that use of perioperative TEE is recommended for all adult open thoracic aortic surgical procedures, i.e., also those involving the aortic arch (70). Also, TEE is indicated in patients with suspected acute aortic syndrome who are unstable and are already intubated (70).

During hybrid and endovascular thoracic aortic procedures, TEE should at least be available. Use of TEE may be considered, e.g., in dissection cases and when general anesthesia is provided, for purposes of procedural and instrumentation guidance, i.e., guidewire placement via the dissected aorta (71), endoleak assessment (71–74) or detection of cannulation injury. The benefit of TEE in these scenarios is less well supported by evidence than for perioperative use. Since some endovascular procedures may be performed under local anesthesia, anesthesia or increased sedation requirements for purposes of TEE monitoring are to be weighed against its incremental diagnostic benefit.

#### **Invasive arterial pressure monitoring**

During endovascular or surgical repair of aortic arch pathology, continuous monitoring of invasive arterial blood pressure is indicated. Selection of the monitoring site should take vessel pathology into account (e.g. dissection, stenosis, fistula, atheroma, anatomical variants), and must not interfere with vascular access and branch vessel manipulation. In endovascular procedures involving the aortic arch, multiple arterial access sites via lower and upper extremities are usually required. The arterial site dedicated to anesthesia monitoring must therefore be chosen carefully in consultation with the performing team, and on an individualized basis.

Open surgical repair of the aortic arch requires periods of occlusion and selective perfusion of supraaortic branches at least temporarily and often sequentially. A single-site arterial line is not sufficient for uninterrupted monitoring of vital organ perfusion pressures. Bilateral invasive radial artery pressure measurement allows monitoring of cerebral perfusion pressure, without interruption during direct subclavian cannulation or during subclavian cross-clamping for cannulation and during repair. When right axillary antegrade cerebral perfusion (RAACP) is performed via a cannulated graft sewn to the artery, simultaneous monitoring of RAACP inflow pressure and resulting left radial pressure is possible. This may provide information about functional integrity of the Circle of Willis (75), and/or run-off blood flow from the LSA to the DTA (DTA). Nevertheless, bilateral radial pressure monitoring is used in aortic arch surgery only by about 50% of surveyed European centers (76).

Additional femoral arterial (FA) pressure monitoring (preferably at the non-surgical or non-dissected FA) allows assessment of efficacy of distal body perfusion before and after hypothermic circulatory arrest (HCA), and to detect post-repair pressure gradients across the arch. Particularly during rewarming from HCA and for several hours after termination of prolonged cardiopulmonary bypass (CPB) runs, radial pressure often underestimates central aortic pressure, which is better approximated by FA pressure. Due to vasodilatory arteriovenous shunting in the distal upper extremities, radial pressure may underestimate central aortic pressure (measured by direct needle transduction) by up to 20 mmHg mean and 35 mmHg systolic pressure (77,78). Overdiagnosis of “vasoplegic syndrome” or “vasodilatory shock”, with inadequate dosing of vasopressor agents, may be avoided by central aortic pressure verification and use of the FA for early postop pressure monitoring.

For surgical repair of the aortic arch, bilateral invasive upper extremity arterial pressure monitoring should therefore be used routinely. In this type of surgery with prolonged CPB and periods of HCA, consideration should also be given to intermittent direct central aortic pressure reference measurement and / or additional FA pressure monitoring.

#### **NIRS-based regional oxygenation monitoring**

Near-infrared spectroscopy (NIRS) of hemoglobin fractions can be used to continuously monitor the balance of oxygen supply and demand in superficial cortical regions of the brain, i.e., by bifrontal NIRS-derived cerebral oximetry (79,80). The potential and the limitations have extensively been studied during carotid endarterectomy where the evidence to define clear cut-off points for the presence of perioperative cerebral ischemia still is limited (81). During aortic arch procedures, cerebral tissue Hemoglobin (Hb) may desaturate for a large variety of reasons, e.g., global or unilateral hypoperfusion (82) or cerebrovenous congestion; aortic or SACP cannula malposition, vessel dissection or malperfusion; systemic hypotension, hypoxemia, hypocapnia, hemodilution, anemia or low cardiac output; insufficient levels of hypothermia or anesthesia; aggressive rewarming (83); or

other causes of regional or global ischemia. If this monitoring modality is used, differential diagnosis and the use of an algorithmic approach to intervention for regional cerebral tissue Hb desaturation is recommended (84,85).

A survey of 144 European cardiac centers found that NIRS oximetry is used in arch surgery by 65% of institutions (76). It also showed that NIRS oximetry has largely replaced invasive jugular bulb oximetry (76,86). An analysis of open arch surgical strategies at 12 large European centers reported NIRS use for neuromonitoring in all centers (87). The limitation remains that uneventful intraoperative bifrontal rSO<sub>2</sub> (regional cerebral oxygen saturation) tracings do not rule out focal cerebral ischemia, which may occur outside the limited field of view of current NIRS devices. Transcranial Doppler monitoring presents another option to monitor changes in cerebral perfusion but is more complex with regard to the setup and the application during aortic arch surgery.

So far there is only low-grade evidence in adult cardiac surgery (85,88–94), and moderate-grade evidence in thoracic aortic surgery that links intraoperative cerebral rSO<sub>2</sub> desaturation to postoperative new neurological morbidity (83,95–97). Nevertheless, with its perceivably favorable risk-benefit ratio, routine use of non-invasive continuous NIRS monitoring during thoracic aortic procedures is increasing (76,87,98–100). For surgical and hybrid repair of aortic arch pathology, NIRS-based continuous monitoring of rSO<sub>2</sub> is recommended in combination with an algorithmic approach to intervention for desaturation events (84,96). Good evidence for a benefit of NIRS monitoring in endovascular arch repair is still lacking. Indications for its use are pragmatically inferred from surgical (carotid, arch) and stroke populations (99,101,102). NIRS-based continuous monitoring of rSO<sub>2</sub> should therefore be considered at an opinion-based level of evidence.

#### **Central nervous system electrophysiological function monitoring**

Electroencephalography (EEG) (raw or more commonly, processed to parametric display) has been widely used in aortic arch surgery to ensure electrical and cerebral metabolic suppression, to a level of complete electro cerebral inactivity (ECI) prior to HCA. This appears helpful in view of the considerable inter-individual variability in cooling efficacy and ischemic risk (103). Cooling time to cortical isoelectricity is not precisely predictable from tympanic or nasopharyngeal temperature trends, since ECI may ensue within a wide range of temperature, i.e. between 27.2° and 12.5° nasopharyngeal temperature (104). The strategy of HCA with hypothermia-induced ECI has produced increasingly good neurological and survival outcomes over time (105), but evidence as to the incremental benefit from EEG monitoring per se remains scarce.

Nowadays, the strategy of open aortic arch surgery increasingly shifts to using moderately HCA (>= 28°C systemic) combined with hypothermic SACP (106–111), with comparably good major outcomes and lower stroke

rates **(109)**. With this strategy, hypothermic EEG silence is no more targeted during cooling, and EEG monitoring refocuses on detection of ischemia and inadequate anesthetic levels as in other surgical fields. Still, the choice of lower core temperatures should be considered for having a sufficient safety margin according to the expected lower body circulatory arrest time

European and German surveys report that EEG is monitored in arch surgery by a third of polled centers (16%-38%) **(76,98)**. Bilateral EEG has been shown anecdotally to indicate inefficacy of SACP during moderately HCA **(112)**. Further evidence is lacking so far that EEG monitoring improves major outcomes of arch surgery with SACP, or of hybrid or endovascular arch repair. Since its incremental benefit in surgical or endovascular repair of aortic arch pathology is established only by opinion and low-grade evidence, EEG or processed EEG monitoring may be considered according to institutional preferences (e.g. use of HCA) and concomitant indications (carotid crossclamping, monitoring of anesthetics effect).

Monitoring of motor (MEP) or somatosensory evoked potentials (SSEP) can be useful in TAA and thoracoabdominal aortic surgery or endovascular repair in order to guide therapy and to allow early intervention in the anesthetized patient **(113–116)**. A metaanalysis confirmed the good performance of MEP monitoring in detecting postoperative paraplegia in thoracic and/ or thoracoabdominal (TAA/TAAA) open repair **(117)**. Both MEP and- less well investigated- SSEP neuromonitoring have been found useful in prevention and prediction of paraplegia **(118,119)**. In a retrospective analysis, MEP has been found useful in simultaneous arch and thoracoabdominal aortic surgery as part of a protocolized brain and spinal cord protection bundle **(120)**. Selective use of MEP and SSEP monitoring in aortic arch surgical or endovascular repair may therefore be considered based on requirements of the individual patient, surgery or procedure, on the urgency of the procedure and institutional resources **(2)**.

During hybrid arch repair, considerations of extracranial cerebrovascular surgery in anesthetized patients apply, while aortic arch debranching is performed without CPB. During this period, monitoring for cerebral ischemia according to institutional preferences (EEG or SSEP and/or NIRS) should be considered **(99,101,121)**. Subsequent thoracic endovascular aortic repair (TEVAR) deployment may compromise spinal cord collateral perfusion. Depending on the extent of coverage and compromise of collateral flow, MEP or SSEP monitoring during this period should be considered in selected patients to assess integrity of spinal cord function **(103)**.

## **Spinal cord perfusion pressure monitoring and lumbar cerebrospinal fluid drainage**

Distal aortic arch repair involving the DTA and use of the FET may compromise the collateral vascular network, and hence perfusion, of the spinal cord. Segmental spinal artery inflow may become impaired depending on flow characteristics in dissection and the extent of coverage by stent-grafts (122,123). Known contributors to spinal cord injury (SCI) are perioperative arterial hypotension, previous abdominal aortic aneurysm (AAA) repair and loss of LSA inflow (124). A systematic review reported a SCI incidence of 5.1% following FET deployment (125). To date, evidence is insufficient for a recommendation to use prophylactic MEP and/or cerebrospinal fluid (CSF) pressure monitoring and drainage in aortic arch repair with the use of FET (125,126). However, the use of lumbar CSF pressure monitoring and drainage may be considered based on individualized risk assessment for spinal cord ischemia (127,128). In situations of delayed SCI, selective secondary insertion of drainage as part of a treatment bundle is recommended (126,129). As imaging is still not able to provide us with a detailed description of intraspinal collateralization, which might be the answer who is at increased risk for SCI, risk prediction models remains approximations such as the collateral network concept and- developed on that basis- the four territory concept (130,131).

CSF drainage management- CSF pressure is measured in mmHg in the majority of settings (since invention of electronic pressure transducers): cm H<sub>2</sub>O and mmHg are not “close in numbers” but enjoy a firm relationship (1 cmH<sub>2</sub>O = 0.735 mmHg). Spinal perfusion pressure (SPP = MAP-CSFP, or -CVP whichever is higher) can only be determined correctly if arterial and CSF pressure transducers are referenced to the same level (phlebostatic axis = right atrial level) and unit of measurement: Hence, mmHg makes more sense, too, although some drainage systems give parallel scales in mmHg and cmH<sub>2</sub>O (e.g. Medtronic Duet® External Drainage & Monitoring System). After placement, a normal CSF opening pressure is 5-18 mmHg, and CSF may be drained to a target CSFP of 10-12 mmHg, as long as there is no SCI.

Some institutions target the normal preoperative opening pressure, measured on catheter placement, as the individual baseline pressure (124) unless there is reason to suspect spinal cord injury. Drainage should always occur slowly, large bolus CSF withdrawals must be avoided. If SCI occurs, reasonable CSF pressure targets are 8-10 mmHg, with limits on “volume” flow at 40 ml/4 hours, although some groups drain even lower to 7 or 5 mmHg, and larger volumes ( $\leq$  20 ml/h) (132). But there is a clear risk (approximately 1%) of overdraining, intracranial hypotension and consecutive brain damage (subdural hematoma or hygroma, intracranial hemorrhage, brain herniation).

## **Multisite Temperature Monitoring**



During CPB, temperature gradients between different monitoring sites (nasopharyngeal, bilateral tympanic, bladder or Recommendation) develop temporarily during cooling and rewarming, and have to be taken into consideration (133). During open aortic arch surgery, monitoring of nasopharyngeal and tympanic temperatures is Recommendationmended to ensure adequate brain cooling, and to prevent cerebral hyperthermia and associated central nervous system (CNS) injury during rewarming (83,134,135). Additionally, bladder core temperature provides the best information available on to protection of the viscerals, renals, lower extremities and finally the spinal cord.

#### **Point-of Care Coagulation Monitoring**

Surgical as well as endovascular aortic arch repair requires reversible anticoagulation with unfractionated heparin (UFH). Although open surgery on CPB carries a substantially higher risk of major blood loss and transfusion, bleeding complications increase morbidity and mortality with either approach. Both procedural anticoagulation and postoperative hemostasis require laboratory monitoring to minimize both hemorrhagic and thrombotic complications. The whole-blood activated clotting time (ACT) test is a functional point-of-care (POC) method, which is recommended to guide UFH anticoagulation, as well as its reversal with protamine and is indicated as a minimum requirement during surgical, hybrid or endovascular aortic arch repair.

ACT is not highly specific for UFH activity, however, and may be confounded by hypothermia, hemodilution, loss of platelets and of coagulation factors (136) all of which typically occur during aortic arch open surgery. Therefore, and in accordance with 2017 EACTS/EACTA (European Association for Cardio-thoracic Anesthesia) Guidelines for Patient Blood Management, heparin management for arch surgery with prolonged CPB and HCA should consider to use quantitative monitoring of circulating UFH concentrations rather than simple serial ACT measurement (137).

Whole-blood viscoelastic coagulation test systems (thromboelastography-TEG rotational thromboelastometry-ROTEM) provide POC analysis of clot generation and stability with short response time. In conjunction with treatment algorithms, they have been shown to be helpful in differential diagnosis and treatment of post-CPB bleeding (138). Moderate-level evidence from trials of elective cardiac surgery with CPB indicates that use of TEG- or ROTEM-guided transfusion strategies may reduce exposure to allogeneic blood products (139–141) and possibly surgical re-exploration for bleeding (137,142,143). In aortic arch open surgery, viscoelastic POC testing should be considered, in conjunction with perioperative treatment algorithms for bleeding patients, in order to reduce allogeneic transfusion exposure and cost.

988 **RECOMMENDATION 10**

989       -   TEE is recommended during all open thoracic aortic surgical procedures. Class I Level B **(70)**

990       -   TEE is recommended in all unstable intubated patients with suspected acute aortic syndrome. Class I

991           Level B **(70)**

992       -   TEE should be available in hybrid and endovascular thoracic aortic procedures. Class IIA Level B **(71)**

993 **RECOMMENDATION 11** During surgery for aortic arch repair,

994       -   bilateral invasive upper extremity arterial pressure monitoring should be considered. Class IIA Level C

995       -   femoral arterial pressure monitoring should be considered. Class IIA Level C

996       -   intermittent direct central aortic pressure reference measurement should be considered. Class IIA Level

997           C

998

999 **RECOMMENDATION 12** During surgery for aortic arch repair, it is recommended to use bilateral NIRS-based

1000 cerebral oximetry combined with an algorithmic approach to intervention for cortical Hb-Desaturation. Class I

1001 Level B **(82–87,95–99,144–146)**

1002

1003 **RECOMMENDATION 13** In situations of delayed SCI, selective secondary insertion of a CSF drainage as part of

1004 a treatment bundle is recommended. Class I Level C

1005

1006 **RECOMMENDATION 14** During surgery for aortic arch repair, multisite temperature monitoring (at a minimum

1007 nasopharyngeal, tympanic, bladder or rectal probe) is indicated. Class I Level B **(83,134,135)**

1008

1009 **RECOMMENDATION 15** During aortic arch surgery, point-of care coagulation monitoring in conjunction with an

1010 algorithmic approach to transfusion of blood products should be considered. Class IIA Level A **(137,140–143)**

1011

## 6) Therapeutic options- Open aortic arch replacement

Open aortic arch replacement involving all three supraaortic branches without the adjunct of either elephant trunk (ET) repair or in combination with the FET technique has become rare (147,148) (Figures 4 and 5). The ET technique should be applied when the FET technique remains debatable. For instance, in large aneurysmal formations, involving several thoracoabdominal (TA) segments and in very small true lumina with the risk of inducing pseudocoarctation), a FET procedure is not recommended.

The ET technique with and without sewing collar solutions is an optimal solution when secondary surgical TA replacement is to be anticipated. The woven polyester is an ideal fabric to be clamped and to be sewn to with a downstream aortic graft for open descending thoracic or TA replacement. On the other hand, the ET can serve as an ideal landing zone for TEVAR extension if the ET is long enough. Therefore, a sufficient length is advisable. A clip at the end of the polyester graft can simplify cannulation during fluoroscopy. Retrograde perfusion of an ET via the femoral artery is not recommended as this might push the ET into the aortic arch and potentially obstruct supraaortic vessels. Therefore antegrade perfusion via the right subclavian/ axillary artery or via side-branch is recommended. In residual dissection after type A repair, the dissection membrane is usually removed as distal as can at least for the length of the ET so that the ET floats in the common proximal lumen.

However, it should be mentioned that the ET portion should be left adequately long to be accessible in zone 4 in order to serve as a platform for either open surgical or endovascular extension. Regarding the level of the descending aortic anastomosis, in parallel to the FET technique, a proximalization of the anastomosis into zone 2 eases accomplishment as well as bleeding control. Additionally, the risk of left laryngeal nerve palsy is reduced. Finally, a double layer running suture or a strip of tissue will reinforce the anastomosis and will reduce the need for correction stitches for hemostasis.

With regard to supraaortic vessels, selective replantation has the advantage of eliminating the largest amount of native tissue thereby potentially reducing the risk for recurrence. A variety of branched grafts is available and should be used according to experience and preference.

**RECOMMENDATION 16** In case of elephant trunk (ET) implantation, the polyester trunk component should be accessible in zone 4. Class I Level C

1047 RECOMMENDATION 17 An anticipative strategy with regard to potential future operations or interventions is  
1048 recommended in any scenario of proximal aortic repair where later secondary distal repair may be needed. Class  
1049 I Level C  
1050  
1051 RECOMMENDATION 18 In case of ET, the distal anastomosis should be considered to be performed in aortic  
1052 arch zone 2 in order to ease accomplishment and to facilitate bleeding control. Class IIA Level C

## 7) Therapeutic options- FET

FET combines the principles of open arch surgery and endovascular DTA repair (**Figure 6**). The extension of arch replacement into the DTA by a separated stent-graft was first introduced clinically by Dr. Masaaki Kato in October 1994. However, it was not until November 1996 that Dr. Kato reported his experience with this technique in 10 patients (**149,150**). The technique has been used in Europe since 2001 (**151,152**).

The technique is called Frozen Elephant Trunk following the development of a combined vascular and stent-graft prosthesis in one fashion (**153**). Similar to the ET technique, a stent-graft is introduced through the opened arch into the DTA enabling the exclusion of distal arch pathologies in one step. The proximal part of the graft is used for conventional arch replacement. The breakthrough for the widespread application of this technique occurred in year 2005 with the development of the first commercially available hybrid prosthesis, the so called E-vita open™, (154). As a tube fabricated vascular graft is invaginated into a stent-graft according to the principle of the modified ET technique (155) and the whole graft is delivered and deployed into the DTA with an endovascular introducer. The FET armamentarium is completed by a branched hybrid graft, so called Thoraflex™, which enables the reimplantation of the supraaortic vessels separately using three prefabricated vascular branches (156). A side graft allows direct cannulation for antegrade distal perfusion during the arch replacement. There are two other commercially available FETs, the Cronus (MicroPort, Shanghai, China) and the J graft (now Frozenix) (Japan Lifeline, Tokyo, Japan) (157,158)

The indication of FET covers all pathologies of the aortic arch, aneurysm and dissection (**159–161**). Different from endovascular aortic repair, the fixation of FET is performed by a circumferential suture, which eliminates the risk of proximal endoleak. The endoluminal sealing of the surgical suture line by the stent-graft improves hemostasis and makes FET ideal to fix a fragile aortic tissue. This combination of surgical suture and endovascular sealing enables the durable exclusion of antegrade false lumen perfusion in acute and chronic aortic dissection as well as aneurysmal cavities without excessive oversizing of the stent-graft. Particularly in acute aortic dissection a progressive false lumen thrombosis in more than 90% followed by shrinkage and positive remodeling has been reported from several studies (**162,163**). The potential exclusion of the downstream aortic pathology occurs predominantly up to the distal end of the stent-graft, so that FET can be applied curatively only in association with the extension of thoracoabdominal aortic disease in many scenarios. Patients with residual aortic pathology beyond the FET remain at risk for secondary treatment. However, shifting the treatment level by the stent-graft to at least a mid-thoracic level facilitates secondary treatment by using the stent-graft as landing

zone for endovascular or as a docking place for open surgical repair. In case of open TA repair, the capability of the stent-graft to be clamped provides an easier surgical access to perform the anastomosis beyond the arch with less necessity of rib resection, HCA and no risk for laryngeal nerve injury **(164,165)**. However, the texture of the fabric of endovascular/ FET devices is by nature thinner and prone to fabric tears when an anastomosis is directly performed to a conventional polyester graft. Therefore, the suture should include the aortic wall as well as good as can.

In case of endovascular reintervention the stent-graft component provides a safe landing zone for distal extension. Thus, FET can be used in type I and II TAAAs as a first stage procedure when primary proximal sealing cannot be achieved adequately by endovascular means. In this case, sizing and length of FET should be planned considering the requirements of the second endovascular procedure in order to avoid excessive mismatch and a multicomponent secondary endovascular intervention. Generally, FET deployment beyond the transition Zone 4-5 provides a safe length for additional stent-graft deployment and easier retrograde access in case of severe aortic tortuosity. However, care has to be taken in order to avoid extensive covering, which is reported to be associated with increased risk for SCI **(166,167)**.

The technique of FET is similar to classic ET and represents major surgery. Sophisticated cannulation and perfusion techniques have been introduced in order to make antegrade selective cerebral perfusion as safe as can, to reduce lower body HCA times to a minimum and to improve organ protection in general. Considering the sealing properties of the stent-graft, the proximalization of FET fixation from Zone 3 to Zone 2 facilitates the distal anastomosis and reduces the duration of lower body HCA as well as the risk for laryngeal nerve injury **(168,169)**. Combination of FET with LSA debranching minimizes the duration of arch repair and allows the perfusion of all 3 arch vessels for additional cerebral and spinal cord protection. The implementation of selective distal perfusion during arch repair using a side graft or balloon cannulas as endoclamp within the FET reduces lower body circulatory arrest times and thereby improves distal organ protection. In addition, selective myocardial perfusion during arch repair ("heart beating" concept) is used to reduce cardioplegic arrest times and to allow more extensive proximal surgical procedures **(170)**.

To secure FET treatment the use of a guide wire, preferably via the FA under angiographic or echocardiographic control may be of help. In aortic dissections, the wire secures FET deployment within the true lumen. In aneurysms, it facilitates the guidance of FET over thrombus formation and aortic tortuosity avoiding debris mobilization and distal embolization. Angioscopy represents an additional intraoperative tool in visualizing the landing zone and endoluminal obstacles and in controlling the deployment downstream **(171)**. Fluoroscopy during FET introduction is usually not needed **but can be helpful**.

1121  
 1122 FET treatment is critical in acute and chronic aortic dissection with completely depended visceral arteries  
 1123 perfusion from false lumen. In these scenarios, preoperative verification of patent communications between  
 1124 lumina is recommended to avoid malperfusion. In connective tissue disease, the use of stent-grafts is  
 1125 controversial and basically discouraged; in any case avoidance of oversizing is recommended. In DTA rupture a  
 1126 safe distal landing zone for definitive sealing is a prerequisite for FET treatment. The TEVAR component of the  
 1127 FET prosthesis cannot be equally interpreted as a “TEVAR-alone” approach in patients with connective tissue  
 1128 disease as the remaining risk of distal stent-graft induced new entry is different in clinical weight and need for  
 1129 correction than a proximal stent-graft induced new entry or in other words- retrograde type A aortic dissection  
 1130 (172,173). Recently, EACTS has formulated recommendations for use of the FET technique (174).

1131  
 1132  
 1133  
 1134 **RECOMMENDATION 19** The FET technique or TEVAR to close the primary entry tear should be considered in  
 1135 patients with acute type A aortic dissection with a primary entry in the distal aortic arch or in the proximal half of  
 1136 the DTA to treat associated malperfusion syndrome or to avoid its postoperative development. Class IIA Level C

1137  
 1138 **RECOMMENDATION 20** The FET technique may be considered for use in patients undergoing surgery for acute  
 1139 type A aortic dissection to prevent mid-term aneurysmal formation in the downstream aorta. Class IIB Level C  
 1140 **(174)(444)**

1141  
 1142 **RECOMMENDATION 21** The FET technique should be considered in patients with complicated acute type B  
 1143 aortic dissection when endovascular interventions are contraindicated. Class IIA Level C **(161,175,176)**

1144  
 1145 **RECOMMENDATION 22** The FET technique should be considered in patients with concomitant distal thoracic  
 1146 and thoraco-abdominal aortic disease that, in a later stage will or is likely to require either surgical or  
 1147 endovascular treatment. Class IIA Level C

**8) Therapeutic options- Transposition (debranching) of supraaortic vessels and TEVAR and the importance of the LSA in spinal cord blood supply**

Hybrid arch repair (or combined vascular and endovascular treatment) is a combination of both open and endovascular procedures aimed to treat aortic arch disease. The core principle behind this treatment relies on endovascular exclusion of the pathology following the creation of an adequate proximal landing zone (in zone 0, 1 and 2) **(1)** by means of supraaortic transposition (debranching) of one (LSA), two (and LCCA) or three (and IA, i.e. total aortic arch debranching) arch vessels **(Figures 7-9)**.

Debranching options are multiple and can be performed by means of anatomical or extra-anatomical revascularization, with extrathoracic or intrathoracic approaches. The techniques presented in literature are pleiomorphic: from aortic patch reimplantation, to branched or simple grafts interposition and autologous transposition **(177)**. Open and endovascular procedures can be performed simultaneously or with a staged approach (open debranching first and endovascular exclusion as a second stage) according to need and preference (178,179). TEVAR in the aortic arch should be performed preferably with a fixed imaging system.

The main potential advantage of the hybrid approach is the avoidance of aortic cross-clamping, HCA, and CPB with the potential risk reduction} in higher risk patients with proximal thoracic aortic pathology (zone 0 proximal neck). For patients at higher risk of stroke open aortic arch surgery remains the best therapeutic option as extensive manipulation during debranching as well as during TEVAR might cause embolization (178,180,181). Patients presenting with distal arch pathology (zone 1 and 2 proximal neck) should be considered for an endovascular approach with prior LSA and/or LCCA revascularization, if anatomically suitable.

The devices employed for aneurysm exclusion are commercially available stent-grafts mostly designed for the treatment of DTA pathology. The Instructions For Use of these devices require deployment in a proximal and distal landing zone (native aorta or pre-existing graft) with a length  $\geq 25\text{mm}$ , measured on the inner curvature, and a diameter  $< 38\text{mm}$ , measured according to manufacturer recommendations (inner/inner vs outer/outer diameter) **(182)**. Application of such devices in patients affected by connective tissue disease is contraindicated unless both landing zones are within a previous surgical/endovascular graft **(183)**. Moreover, at least one adequate ( $> 7\text{mm}$ ) access vessel is required for successful stent-graft insertion, and the aortic lumen characteristics should be taken into consideration to decrease the risk of embolization during advancement of the device in the aortic arch (e.g. shaggy aortas, floating thrombi, severe calcifications) **(184)**. Possible limitations of the hybrid approach are the lack of inflow vessel for debranching (i.e. calcific/aneurysmatic ascending aorta), and the presence of unsuitable landing zones length/diameter or narrow access vessels, inadequate for stent-graft introduction. Open repair



should be considered in these cases as well as in cases at high risk of retrograde dissection (ascending aorta>38mm, bicuspid aortic valve, arch abnormalities, lost sinutubular junction, extended ascending aortic length).

Furthermore, the hybrid approach carries risk of SCI due to the covered length of the DTA. For this reason, CSF drainage should be employed in patients with increased risk (e.g. previous aortic surgery, occluded hypogastric/subclavian arteries) (188–190. Also in hybrid procedures, current literature supports centralization in centers with adequate volume and expertise (7).

#### **Importance of the LSA in spinal cord blood supply-**

The main reason for prophylactic LSA revascularization prior to TEVAR is maintaining posterior cerebellar perfusion as well as maintaining upper inflow into the anterior spinal artery and thereby spinal cord. There is convincing evidence that the combination of LSA occlusion and extensive coverage of thoracic segmental arteries by TEVAR are associated with increased risk of SCI which is significantly lower when the LSA is preserved. This becomes clear when the collateral network concept and consecutively the four territory concept is conceptually applied (130,131,185–187).

**RECOMMENDATION 23** TEVAR in zone 0 after previous debranching may be considered in patients unfit for open repair and suitable anatomy. Class IIB Level B (180,191)

**RECOMMENDATION 24** TEVAR in zone 1 and 2 should be considered in patients with suitable anatomy Class IIA Level B (4)

**RECOMMENDATION 25** Stent-graft deployment is not recommended in patients with a proximal and/or distal landing zone length less than 25mm or a maximum diameter of more than 38mm. Class III Level B (4,191)

**RECOMMENDATION 26** Zone 0-2 TEVAR is not recommended in patients with connective tissue disease if the proximal landing zone is in native aortic tissue. Class III Level C

**RECOMMENDATION 27** Open aortic arch repair should be considered in patients with concomitant aortic valve pathology or at high risk for retrograde type A aortic dissection (ascending aorta>38mm, bicuspid aortic valve, arch abnormalities, lost sinutubular junction, extensive ascending aortic length). Class IIA Level B (175,191)

1217

1218 RECOMMENDATION 28 In elective TEVAR in zone 0,1,2, preventive left subclavian artery revascularization  
1219 should be considered to reduce the risk of neurological complications such as stroke and spinal cord ischemia

1220 Class IIA Level B **(152,171)**

1221

1222 RECOMMENDATION 29 Hybrid aortic arch repair should be centralized in centers with adequate volume and  
1223 expertise in both open and endovascular surgery. Class I Level C

1224

## 9) Therapeutic options- Total endovascular repair

The development of new endovascular techniques to treat aortic arch aneurysms has mitigated the risks associated with open surgery and offers repair to patients that historically cannot undergo open repair. Early experience using external branch endo-grafts suffered from high stroke rates, and were not adopted in the global market (171-174). The subsequent development of arch endo-grafts with specific delivery systems, preloaded fenestrations and inner-branches in recent years has improved results to a level that endovascular arch repair has today become a viable option for patients with increased risk for open repair (Figure 10).

In contrast to more stable segments of the aorta, where endovascular treatment has become the standard of care, the ascending aorta is characterized by high velocity and consequent shear stresses, four-dimensional pulsatile and rotational movements during the cardiac and respiratory cycles, and the proximity of the coronary ostia and aortic valve. Endovascular arch repair requires a stable proximal landing zone within a surgical graft or native ascending aorta with a diameter of 38mm or less. Larger diameters are prone to retrograde dissection and thus should be avoided (196). The proximal sealing-zone should preferably have a length of 30mm or more measured at the inner curvature that is free of excess calcification and thrombus, and angulation >60°.

Stroke remains a major concern during endovascular arch repair, with rates between 0 and 14% (176-181). The mechanism of stroke includes solid emboli released by manipulation in the arch, air emboli released from the delivery system, and coverage of the target vessels (193). To minimize the stroke risk, temporary carotid artery occlusion, filter placement and carbondioxide flushing of the delivery-system has been proposed (193,194,200).

For endovascular aortic arch repair, there are two general graft designs: fenestrated and branched arch endo-grafts. Both designs are currently available as custom made devices only, so manufacturing time of between 4 and 8 weeks precludes its use in urgent and emergency situations. The current two inner branches design may however become a future platform of an "off the shelf" branched stent-graft that will be used in emergency situations.

Fenestrated arch endo-grafts can incorporate multiple fenestrations or a combination of fenestrations and scallops. Graft apposition to the aortic wall at the level of the fenestrations is required for endovascular seal. The sealing zone therefore is usually in the mid-arch at the level of the branch-vessels. Due to the distance from the

femoral access vessels and the curvature of the arch, rotation of the fenestrated graft cannot be controlled, so precision of placement relies on meticulous preoperative planning and the use of precurved delivery systems and pre-loaded catheters that allow wires to be passed via these catheters and snared from upper extremity access.

The largest cohort of fenestrated arch repair from Japan used a pre-curved fenestrated stent-graft (Najuta-graft) without preloaded wires in 363 patients with a landing zone in the ascending aorta and reported a 1.6% 30d-mortality and 1.8% stroke rate (201). This system does not use bridging stents to fixate the fenestrations at the target-vessel ostia. The Zenith fenestrated arch endo-graft (Cook Medical, Brisbane, Australia) uses a preloaded wire system combining usually a fenestration and a scallop using a covered bridging stent to fixate the fenestration to the left LCCA or LSA as target vessel. Small published series representing early experience have reported mortality up to 20% and stroke rate up to 14% (192,202). The Relay scalloped endo-graft (Terumo Aortic) does not include fenestrations, preloaded catheters or the use of covered stents for the target-vessels and is mainly used for zone 2 and 3. In a single reported small series mortality was 5% and stroke rate 14% (197). **Stroke remains a major concern in any kind of open or endovascular aortic arch treatment strategy and can be seen as the major important challenge to address in the years to come.**

Branched arch endo-grafts currently include antegrade or retrograde internal side-branches along the outer curve of the stent-graft. The two currently available platforms in Europe aiming at seal in the ascending aorta both use 2 antegrade inner-branches to be connected to the IA and to the LCCA while the LSA is usually debranched in a staged procedure (193). This design requires less precision in placement compared to fenestrated arch endo-grafts as distance between the branch openings and the target-vessels allows for continued perfusion of the supra-aortic vessels after main-graft deployment and a simplified catheterization of the inner-branches.

The Zenith branched arch endo-graft (Cook Medical, Brisbane, Australia) includes a staged proximal release mechanism. Early experience has been reported with no 30d mortality and a stroke rate of 11% (199). The Relay branched arch endo-graft (Bolton Medical, Barcelona, Spain) is built using two parallel inner-branches on the Relay NBS platform using a proximal tip-capture. Early experience in a small series collecting global experience showed a 7% mortality and a 7% disabling stroke rate (203).

Branched endovascular arch repair is today increasingly used in patients after previous open ascending repair for type A aortic dissection. The presence of a prosthetic graft in the ascending aorta acts as a favorable proximal landing zone for an arch endo-graft, excluding risk of retrograde dissection and > 70% of patients have a proximal landing zone in the previous ascending aortic graft suitable for branched endovascular arch repair (204).

1294 The TAG® single sidebranch endo-graft (Gore® Medical, Flagstaff, USA) and the Valiant™ Mona LSA single  
1295 sidebranch stent-graft (Medtronic Inc, Santa Rosa, USA) are arch endo-grafts with a single sidebranch aiming at  
1296 preservation of the LSA in Zone 2 TEVAR **(205)**. Both endo-grafts are currently used in limited number of centers  
1297 with no clinical data published so far.

1298

1299 At the present time, with careful patient selection and operator experience, early use of this technology presents  
1300 an alternative to open aortic arch repair or conservative therapy, respectively.

1301

1302

1303 **RECOMMENDATION 30** Endovascular aortic arch repair in zone 0 should be considered in patients unfit for open  
1304 surgery and with a suitable anatomy. Class IIA Level B **(199,203)**

1305

1306 **RECOMMENDATION 31** It is recommended that endovascular aortic arch repair is performed in centers with  
1307 adequate volume and expertise of open and endovascular arch repair. Class I Level C

1308

1309 **RECOMMENDATION 32** In any open proximal thoracic aortic surgery, ascending/ hemiarch replacement has to  
1310 be extensive and short ascending grafts should be avoided for preventing disease progression and for  
1311 anticipating future endovascular modular distal extension. Class I Level C

1312

1313

## 10) Therapeutic options- Alternative approaches

Alternative approach to aortic arch pathologies are endovascular techniques applying the chimney graft (CG), the periscope and sandwich technique (summarized as parallel grafts) and in-situ fenestration. Parallel grafts are bare or covered stents deployed into one or more supraaortic vessels parallel to the main aortic arch stent-graft. This allows extending the sealing zone of the aortic stent graft beyond the origin of the respective supraaortic vessel. One of the first reported parallel grafts in the literature was used in 2003 (206) in a patient undergoing EVAR to secure a renal artery in a patient with a very short proximal landing zone. The first parallel graft used in aortic arch treatment was reported 2 years later (207). There are several modifications of the parallel graft technology. The standard parallel graft is proximally oriented and allows antegrade flow up to an aortic branch. The periscope parallel graft is distally oriented and blood flow is retrograde. The sandwich technique includes an aortic stent graft deployed first as an artificial landing zone to implant the parallel grafts. After parallel graft implantation another aortic stent-graft is deployed to exclude the entire pathology. The parallel grafts are located between both aortic stent-grafts. Furthermore, parallel grafts can be used only to compress the graft edge to secure the flow into the vessel where the parallel graft was implanted. Parallel graft are used as a bailout when target vessels are incidentally covered to allow very aggressive stent graft placement in case of short landing zones 2 and 3.

There are several advantages of parallel graft techniques when compared with fenestrated or branched stent grafts. First of all, parallel grafts are available off the shelf. Fenestrated and branched stent grafts are mostly customized and usually manufacture time takes 1-3 months. They are clearly not an option in patients requiring emergent or urgent aortic arch repair. Second, parallel grafts are less expensive than fenestrated and branched stent grafts. Furthermore, there is a large experience of visceral CGs available in the literature with acceptable results especially in patients requiring urgent aortic repair (208). However, the literature on supraaortic parallel grafts is scarce. Results of visceral parallel grafts are most probably not representative for expected results of supraaortic parallel grafts.

Parallel graft techniques carry a risk of endoleak Type I due to the so-called gutters, which are channels between the parallel graft and the main aortic stent graft. Those gutters are per definition inevitable, however not all of them lead to endoleaks detectable in CTA. Even thrombosed lesions can still remain under pressure, if there are gaps in the sealing zone. It may lead to endotension which is defined as pressure within the aneurysm sac without evidence of endoleak as the cause. Endotension raises the risk of aneurysm rupture (209). Gutters caused by parallel grafts are specifically relevant if pathology at the outer curvature is treated such as in most cases of Type B aortic dissection, where gutters may cause Type 1A endoleak. If pathologies affecting the inner

curvature are treated, gutters caused by parallel grafts on the outer curvature are less prone to cause type 1A endoleak.

Furthermore, stent-grafts were not designed for the parallel graft approach. The radial force, elasticity, shape and even length of currently used stent grafts in parallel graft cases are not optimal. There are no covered stent grafts dedicated for parallel graft techniques. Additionally, numerical studies suggest worse hemodynamic performances in parallel graft models when compared with surgical or hybrid arch repair models **(210)**. Finally, to avoid endoleak type 1 aortic stentgraft oversizing is necessary and for larger aortas aortic stent-graft with appropriate diameter are not available **(211)**.

There are several reports on parallel graft in treatment of aortic arch or proximal DTA. The largest multicenter series include up to 95 patients **(212)**. The 30-day mortality ranges between 0 and 29% (including elective and emergency cases **(213–215)**) The overall early patency rate of parallel grafts ranges between 92 and 100% **(178–180)**. Early endoleak type I was reported in a meta-analysis of 314 cases at the level of 11% (range, 0-44%) **(216)**. Forty-five percent of early endoleaks in this report sealed spontaneously. The follow-up in currently available reports ranges between 1 and 30 months **(212–216)**. There are no long-term follow-up data in these patients. The number of re-interventions is provided in most reports, however in the vast majority of reports there are no data on number of patients who require aortic arch repair due to the failure of parallel grafts. Finally and most importantly in vast majority of reports there are no data on sac dynamics at follow-up.

Parallel graft technology is a useful treatment option in patients requiring emergent or urgent aortic arch repair. Parallel grafts can also be used as a bailout technique in case of accidental covering of a supraaortic vessel. Parallel grafts should be avoided in elective cases with anatomy suitable for branched or fenestrated devices or open surgery until more data with better quality than currently available exists.

In-situ fenestration of standard stent grafts is another option to extend the proximal landing zone by covering the supraaortic branches and performing a fenestration via a retrograde access in vivo **(217,218)**. Graft perforations can be performed by laser or mechanical means. This technique is new and long-term data in human are missing. Currently in-situ fenestration is an off-label procedure that can be used only as an emergent bailout technique or in the setting of investigational studies. Recent work demonstrates that both laser and mechanical in-situ fenestration create substantial damage to all available stent-graft fabrics **(219)**.

The multilayer (or flow modulator) technique has recently been advocated for the treatment of various thoracic and abdominal aortic pathology including the aortic arch. The principle of the technique is formed by a self-

1382 expanding multi-layered stent constructed of cobalt alloy wires interconnected in five layers. Thereby, blood flow  
1383 through the stent is laminated reducing turbulence in the aneurysmal sac leading to sac thrombosis. Conflicting  
1384 evidence regarding the mechanisms and efficacy currently remain unsolved (220–224)

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1386 RECOMMENDATION 33 The parallel graft technique should be considered in urgent TEVAR procedures  
1387 requiring seal in landing zones 0-2 without adequate options for open surgery or supraaortic debranching and as  
1388 a bail out strategy in cases where unintended obstruction of a supra aortic vessel occurred during TEVAR. Class  
1389 IIA Level C

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1391 RECOMMENDATION 34 The parallel graft technique is not recommended as a routine strategy in preserving flow  
1392 to major supraaortic branches in Zone 0-2 if other strategies (open surgery, branched/fenestrated stent-grafts) are  
1393 available. Class III Level C

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1395 RECOMMENDATION 35 The Multilayer technique is not recommended for the treatment of any kind of aortic  
1396 arch pathology. Class III Level C

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1400

11) Ten bullet points when to choose what kind of approach

Factors favoring one or the other approach	Endovascular repair	Open repair
Previous CABG with patent IMA graft at risk at resternotomy	V	X
Poor LV- or RV-function	V	X
Poor pulmonary function	V	X
Poor liver function	V	X
Connective tissue disorder patients with landing zones in native tissue	X	V
Access vessels (femoral and iliac) diameter < 7mm	X	V
Native ascending aorta diameter > 38mm	X	V
Valvular heart disease necessitating concomitant repair	X	V
Previous mechanical aortic valve replacement	X	V
Prosthetic ascending aorta short or kinked	X	V

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1404 V favors

1405 X discourages

## 12) Rare pathologies

### a) Thrombus

Aortic thrombus is a rare entity (225). The aortic arch and the DTA have been recognized as predilection sites for aortic thrombus (226). Aortic arch thrombus bears the risk of life-threatening stroke and peripheral embolization (227). The thrombus morphology should be taken into consideration distinguishing mobile (i.e. floating, bulging into the lumen) from stationary (mural lining) thrombus. Symptomatic patients (i.e. ischaemia due to embolization: stroke, limb ischaemia, visceral or renal ischaemia) often require urgent treatment and in asymptomatic patients the diagnosis is mostly a chance finding in imaging studies performed for other reasons. There is a high prevalence of hypercoagulation and hematologic disorders including malignancy in patients with aortic thrombus (226). This has to be considered when establishing individual treatment strategies. Other possible sources of embolization have to be ruled out preoperatively in symptomatic patients. Treatment options include conservative management (anticoagulation) and surgery (thrombectomy, local resection of attachment site, aortic arch replacement or debranching and thoracic endovascular repair). However, endovascular treatment requires an adequate landing zone in the ascending and DTA. Furthermore, guidewire and stent-graft manipulation in the thrombotic aortic arch bear an additional risk for embolization. A hybrid approach with supra-aortic debranching and antegrade stent-graft implantation has been reported (228). A recently published case series reported excellent outcome with regard to survival and freedom from recurrence of thrombus formation with surgical thrombectomy (227). The value of minimally invasive approaches including trans-arterial balloon thrombectomy or catheter based percutaneous thrombus aspiration remains unclear. Follow-up imaging is recommended in patients under conservative treatment to assess for thrombus dissolution.

**RECOMMENDATION 36** Surgical treatment should be considered in symptomatic patients with floating aortic arch thrombus (IIA C)

**RECOMMENDATION 37** Surgical treatment may be considered in symptomatic patients with extensive stationary (mural lining) aortic arch thrombus (IIB C)

## **b) Aberrant subclavian artery and Kommerell's diverticulum**

The prevalence of aberrant subclavian artery and Kommerell's diverticulum is 0.4-2.3 % (229). Anatomically, the aberrant subclavian artery passes in 80 % posterior to the oesophagus, in 15 % between the oesophagus and the trachea, and in 5 % anterior to the trachea (229). Symptomatic patients suffering from dysphagia, dyspnoea, coughing, chest pain, aspiration, or recurrent pulmonary infection represent only 5 %. Asymptomatic patients can be managed conservatively. Aneurysmatic aberrant subclavian arteries  $\geq 3$  cm in diameter and Kommerell's diverticula with a diameter  $\geq 5.5$  should be considered for repair due to their risk of rupture and dissection. But, actual size measurement of the Kommerell's diverticulum is highly controversial with no clear consensus. Tanaka et al. recommend to measure Kommerell's diverticulum from the wall next to the trachea to the opposite aortic wall or from the tip of the diverticulum to the opposite aortic wall. Additionally, they measure the subclavian artery diameter at its orifice (230). Operative treatment modalities include resection and ligation of the symptomatic or aneurysmatic aberrant subclavian artery to release compression (important in symptomatic patients) and subclavian-carotid transposition or bypass to re-establish arterial circulation to the right arm. Resection of the offspring of the aberrant subclavian artery is not necessary in asymptomatic patients. Kommerell's diverticulum can be treated by stent-graft implantation or DTA replacement. TEVAR might be challenging due to steep arches; often present in these patients.

**RECOMMENDATION 38** An aneurysmatic subclavian artery ( $\geq 3$  cm) and/or Kommerell's diverticulum ( $\geq 5.5$  cm) should be considered for repair. Class IIA Level C

**RECOMMENDATION 39** Treatment should be performed in symptomatic patients with aberrant subclavian artery and Kommerell's diverticulum. Class I Level C

## **c) Trauma**

Aortic injury is highly lethal representing the second most common cause of death in blunt trauma after brain injury. A lesion at the aortic isthmus in loco typico is present in up to 90% of deceleration trauma patients admitted to hospital alive. An autopsy study of 242 fatal blunt aortic injuries showed that isthmus lesions represented 58 % and aortic arch lesions were rare (3 %) (231). Iatrogenic lesions associated with catheter manipulation in the arch is another possible source of trauma. Timing of repair conforms to the extent of the lesion. Classification of traumatic aortic injury according to Azizzadeh et al includes four grades of lesions: I intimal flap, II intramural haematoma, III pseudoaneurysm, and IV rupture (232). Whereas grade I and II lesions permit conservative management with serial imaging controls, grades III-IV should be repaired. Operative treatment modalities include a hybrid approach with supra-aortic debranching and stent-graft implantation or aortic arch replacement.

1475 Endovascular management is preferred when feasible. Timing, type and extent of treatment also strongly depend  
1476 on concomitant injuries (e.g. traumatic brain injury).

1477

#### 1478 **d) Infection**

1479 Infection of the native aorta or, more often, of an aortic graft encompasses considerable morbidity and mortality.  
1480 For diagnostic purposes a positron emission tomography (PET) scan may add value to differentiate general  
1481 inflammation (e.g. postoperatively) from infection. However, metabolic activity on PET-CT is only a minor  
1482 criterium. The MAGIC (Management of Aortic Graft Infection Collaboration) criteria offer support in the diagnosis  
1483 of aortic graft infection (233). Summarizing, the diagnosis of native aortic or prosthetic aortic infection includes  
1484 clinical/ surgical, radiological, and laboratory data (233).

1485

1486 Operative treatment modalities include removal of the infected material, local debridement and in-situ aortic  
1487 reconstruction. Conservative treatment may be considered in selected cases (234). TEVAR as emergency  
1488 therapy despite suspected aortic infection is feasible and may well serve as a definite treatment option in selected  
1489 cases (235).

1490

1491 Specific antibiotic and antimycotic treatment according to microbiological analyses has to be established for all  
1492 patients. The appropriate type of material for aortic reconstruction is under discussion: prosthetic (plain, antibiotics  
1493 or silver coated) or biologic (homograft, autologous veins, xenopericardial material) grafts are available. The  
1494 required treatment urgency has an influence on preoperative diagnostic features (imaging and microbiological  
1495 sampling) and the availability of the specific replacement material. Xenopericardial material (self-made tube  
1496 grafts) due to permanent off the shelf availability, ease of handling, and good clinical results is favoured  
1497 **(236,237)**. In addition, antibiotic therapy may be withdrawn in many cases during follow-up which is the exception  
1498 in patients after alloplastic replacement.

1499

1500 **RECOMMENDATION 40 Removal of the infected vessel or prosthetic material, local debridement and in-situ**  
1501 **aortic reconstruction using biological material should be considered in infections of the native aortic arch or aortic**  
1502 **arch graft. Class IIA Level C**

1503

1504 **RECOMMENDATION 41 Endovascular repair may be considered for bridging purposes or definite treatment in**  
1505 **inoperable patients in infections of the native aortic arch or aortic arch graft concomitant to antiinfectious**  
1506 **therapy. Class IIB Level C**

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## **Aortitis of the aortic arch**

Immune-mediated vasculitis represents a frequent and possibly organ- or life-threatening disease in rheumatology's every-day practice.

Large vessel vasculitis is the most frequent cause of vasculitis encountered mostly in either young females known as Takayasu's arteritis (TAK) or in people over the age of 50 years known as giant cell arteritis (GCA) (238). Both entities share a possible affection of the aortic arch with mostly late detection, the risk of ongoing inflammation leading to stenosis and dilatation, finally encountering the risk of aortic rupture.

## **Giant cell arteritis**

GCA might present with a sudden onset of temporal headache, malaise with signs of a systemic inflammatory response syndrome (SIRS) of unknown origin, weakness of shoulder and hip girdle and weight loss. Formerly known as Horton's disease and representing a segmental vasculitic affection of the temporal arteries novel diagnostic methods allowed to broaden our understanding of the disease. Meanwhile, CTA as well as MRA and / or PET-CT are able to detect additional vasculitic affections of the aorta that are mainly located in the region of the aortic arch, DTA and in part abdominal aortic or iliac sections.

## **Diagnostic approach**

Further diagnostic evaluation of possible aortic affections is reasonable in order not to miss concomitant large vessel vasculitis (239). Glucocorticoid (GC) treatment should be withheld until after the procedure if medically justifiable: 3 to 5 days after start of GC treatment vessel wall signals mostly disappear resulting in negative results despite underlying inflammation (239,240).

## **Therapy**

In case of temporal arteritis immediate initiation of therapy is warranted in fear of further vasculitic affection of the vasculature supplying the optical nerve with a possibly rapid onset of mostly irreversible blindness. In case of additional or isolated large vessel vasculitis rapid reduction of vessel wall inflammation is supposed to reduce further sequelae. GCs hereby still represent the mainstay of therapy. Current investigations demonstrated IL (Interleukin)-6 as being mainly involved in orchestrating disease onset as well as the course of disease: meanwhile, therapeutic strategies targeting IL-6 and its specific receptor have proven beneficial in inducing and maintaining remission (241,242).

1542

1543 **Complications and outlook**

1544 Rupture of the aorta and/or its associated branches appears to represent a rare complication yet true incidences  
1545 are difficult to depict as e.g. “silent” GCA will not routinely be followed clinically and/or radiographically.  
1546 Furthermore, the former routine for histological evaluation of the resected vasculature in order to prove  
1547 immunologically driven inflammation has unfortunately lost importance. Follow-up of patients focusses on clinical  
1548 and serological signs of relapse and/or remission. For the time being radiographic diagnostics are not reliable for  
1549 determining ongoing or recurrent vascular inflammation: despite clinical and serological remission persistent MRA  
1550 signals within the vessel wall might represent either persistent low disease activity or formation of new  
1551 vasculature or even display some kind of vascular repair. Nevertheless, MRA and/or PET-CT might prove useful  
1552 in the early detection of vascular damage and should therefore be performed repeatedly.

1553

1554 **Takayasu’s arteritis**

1555 TAK manifestations are rarely suspicious of an underlying immune-mediated vascular process but rather point to  
1556 vascular damage after the development of stenosis. As mostly supra-aortic branches of the aortic arch are  
1557 affected patients often present with pulselessness of upper extremities, arm claudication, dizziness or suspicion of  
1558 cerebral ischemia.

1559

1560 **Diagnostic approach**

1561 The diagnostic procedure comprises the same imaging methods as in GCA with US being relevant for supraaortic  
1562 branches, and CTA, MRA and PET-CT being reserved for screening of remaining aortic involvements. Active  
1563 lesions are more probably being detected in phases of serological inflammation.

1564

1565 **Therapeutic approach**

1566 As in GCA, initial therapy comprises the use of GCs aiming at induction of remission. Therapeutic strategies for  
1567 maintenance of remission are not well characterized within this rare disease. Unspecific immunosuppression  
1568 targeting the involved lymphocytic subgroups by using e.g. azathioprine or methotrexate have empirically proven  
1569 beneficial. Therapeutic strategies aiming at TNF (Tumor necrosis factor)-alpha and anti IL-6 yield positive results  
1570 and suggest further adaptations for future therapies (243,244).

1571

1572 **Complications**

1573 Vascular reconstruction might be ~~demanded~~ indicated in the late phase of the disease when symptomatic  
1574 stenoses occur leading to reduced perfusion in the connected arterial segment. Vascular interventions as e.g.

1575 dilatations appear of only little benefit due to the inflammatory nature of the disease with prompt re-stenoses  
1576 occurring quite frequently. Stenting and/or vascular repair have proven more beneficial.

1577 **Conclusion**

1578 Overall, suspicion of large vessel vasculitis due to autoimmune pathophysiology should be considered in either  
1579 young females presenting with mostly late complications of upper extremity claudication and supraaortic  
1580 malperfusion (TAK) and in people ~~over the age of~~ **older than** 50 years with sudden onset of an inflammatory  
1581 syndrome of unknown origin, temporal headache and constitutional symptoms (GCA). Prompt diagnosis and  
1582 therapy especially in GCA will help to minimize the initial risk of permanent loss of vision and to reduce the  
1583 occurrence of long term vascular complications.

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**13) Durability and reporting standards/ quality indicators**

While reporting standards in classical adult cardiac or vascular surgery have widely been established, there is work to do in the aortic sector in particular when it comes to treatment of the aortic arch. As long as no preoperative risk stratification score for aortic disease has been established, currently available risk score systems like STS-PROM (67) or ES I and II (68) may help in predicting risk with their known limitations when applied to patients with aortic disease. However, the main advantage when using them in their current form is the potential comparability between studies where currently there is no least common denominator available.

The results of endovascular repair should be reported according to current SVS (Society of Vascular Surgery) guidelines that consider both technical and clinical endpoints in order to evaluate the performance of the devices combined to the clinical outcomes of their application. Clinical outcomes for aortic arch treatment should clearly include 30 day mortality as well as neurological outcomes (stroke and spinal cord ischemia). Moreover, the completeness of follow-up information is of paramount importance and cannot be overemphasized (245). Neurological outcomes should be reported according to current recommendations (246).Currently, there is no robust evidence to recommend minimum case-loads for aortic arch procedures both open and endo neither for centers nor for individual physicians but a clear volume-outcome correlation like in many other cardiovascular procedures supports centralization and specialization (16,87,247).



#### 14) Gaps in Evidence

Supportive evidence level in above recommendations for the management of aortic arch diseases is mostly “C”, for several reasons. The patient population requiring aortic arch procedures is small compared to other cardiovascular patient populations, although growing. Caseload is low in many centers, and published series tend to be small in numbers. Also, there is much heterogeneity in presentations, patients, and treatment approaches. In particular, therapies in the area of aortic arch pathologies are driven by rapid innovations in technology as well as by institutional preference. Therefore it is very clear that close international scientific and clinical collaboration will be required to solve these issues.

The following unmet needs and gaps in evidence are identified, as a topic of future clinical research in the field:

**An increase of evidence in the pathophysiology and in the prevention of perioperative stroke**

- **An increase of evidence in selecting the best treatment option in patients with acute and chronic aortic arch disease**

- A need for further international standardization of terminology
- A need for standardized surveillance and for follow-up after treatment
- A need to develop prospectively maintained, large multicentric clinical databases for aortic arch pathologies.

This is in recognition of the shortcomings of current Cardiovascular Surgical Risk Scoring Systems in this field (248). As an initiative, the STS Task Force on Aortic Surgery has already developed new sections pertaining to aortic root and thoracic aortic surgery to reflect technical advances in open and endovascular aortic procedures (69). An exemplary set of pertinent variables is given in the STS Aorta Surgery Worksheet V2.9 (249).

- Data-driven development and continuous adaptation of dedicated CPM for Aortic Arch Repair
- Accrual of more evidence on effects of caseload and centralization of care on outcome of aortic arch repair
- A need to address frailty (250–252) and gender differences in outcome research
- A need to define differences in the risk of acute dissection among genetically mediated aortic disease syndromes
- In type A dissection, to better define the extent of index surgery
- Improving the evidence for measures to reduce lower body circulatory arrest time and for selective myocardial perfusion during open aortic arch repair

1643 - To resolve the controversy upon the use of stent-grafts in connective tissue disease

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## **18) Figure legends**

2669 **Figure 1-** Definition of attachment zones also known as Ishimaru zones

2670 **Figure 2-** Aortic arch configurations

2671 **Figure 3-** Definitions of aortic dissections

2672 **Figure 4-** Aortic arch replacement using either the island technique or the selective reimplantation technique

2673 **Figure 5-** Aortic arch replacement using the elephant trunk technique with the descending anastomosis in zone 2

2674 **Figure 6-** Aortic arch replacement using the frozen elephant trunk technique with the descending anastomosis in  
2675 zone 2

2676 **Figure 7-** Subclavian-to-carotid transposition

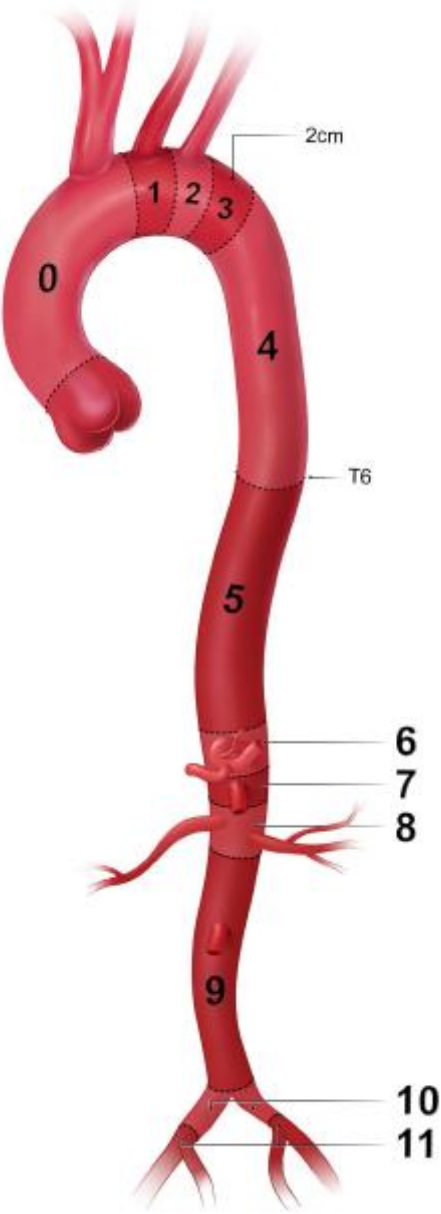
2677 **Figure 8-** Subclavian-to-carotid bypass and Amplatzer plug insertion in the proximal left subclavian artery

2678 **Figure 9-** Autologous double transposition of the supraaortic branches

2679 **Figure 10-** Total endovascular aortic arch repair using the double branch technique

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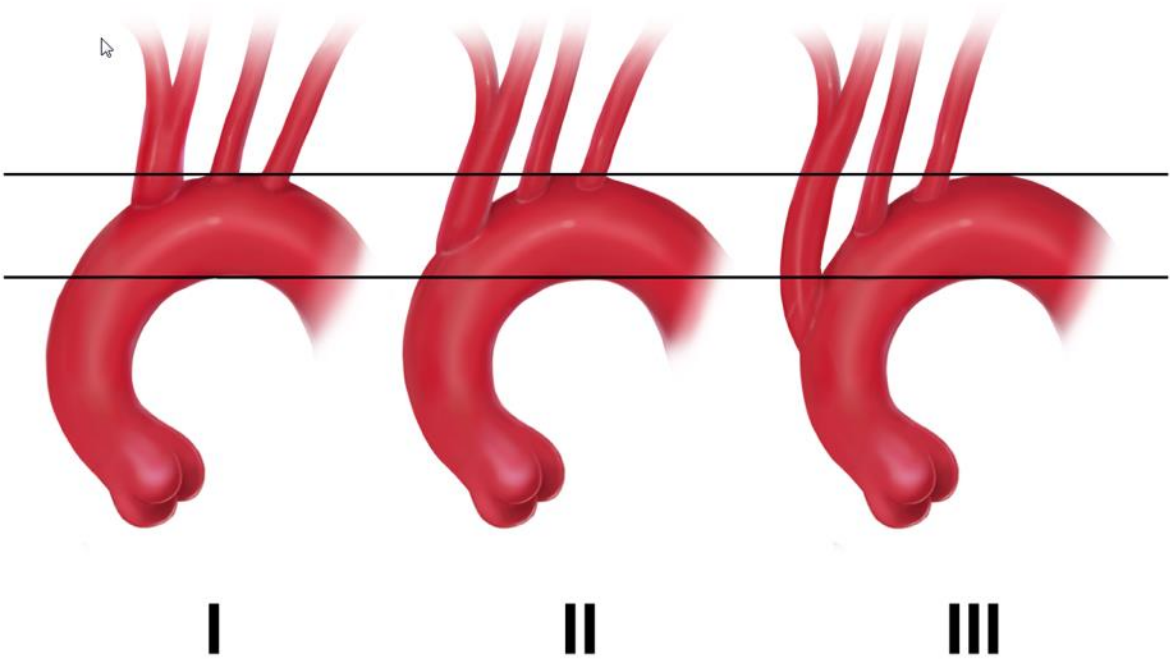
2681  
2682 **Figure 1**



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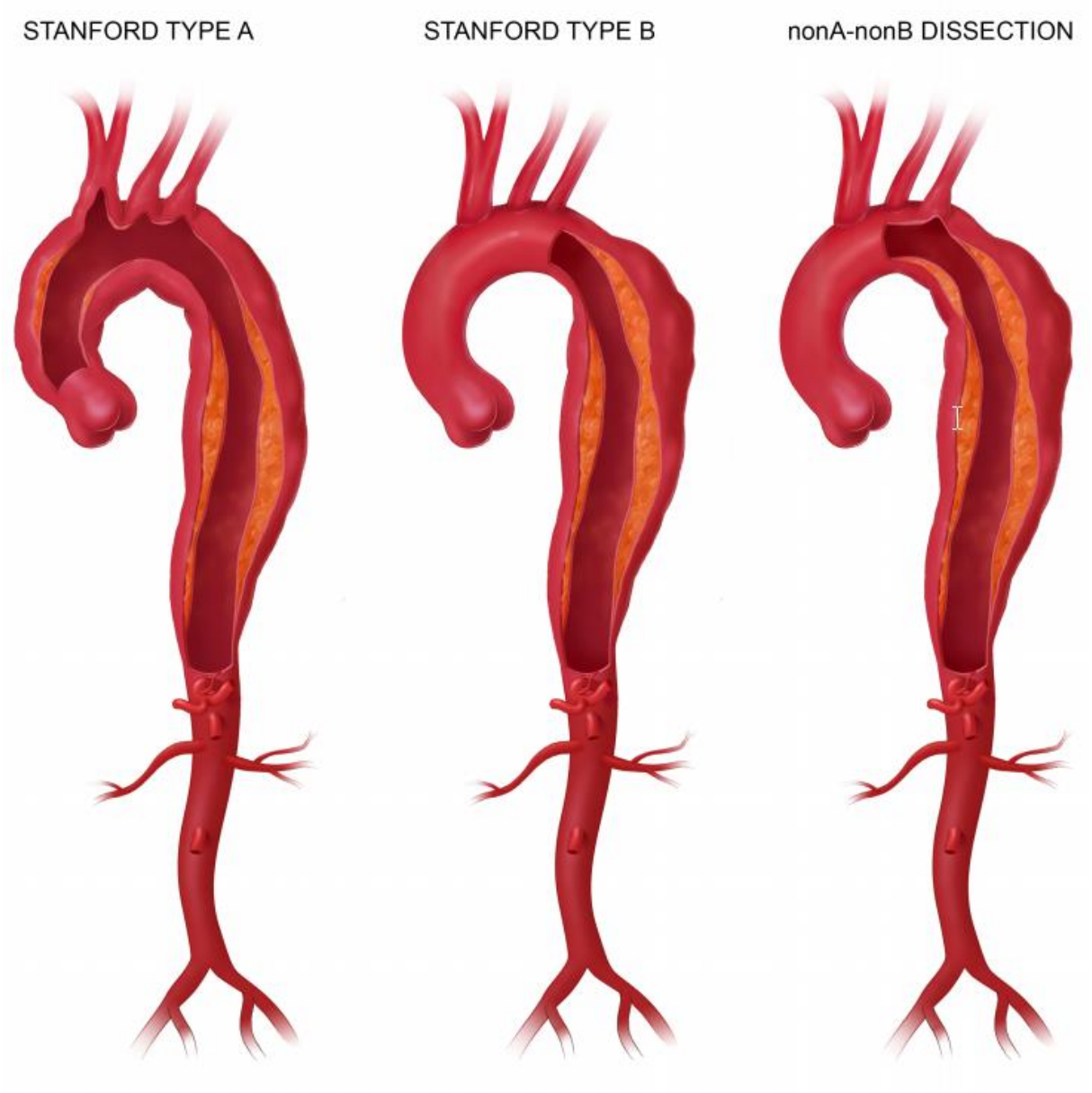
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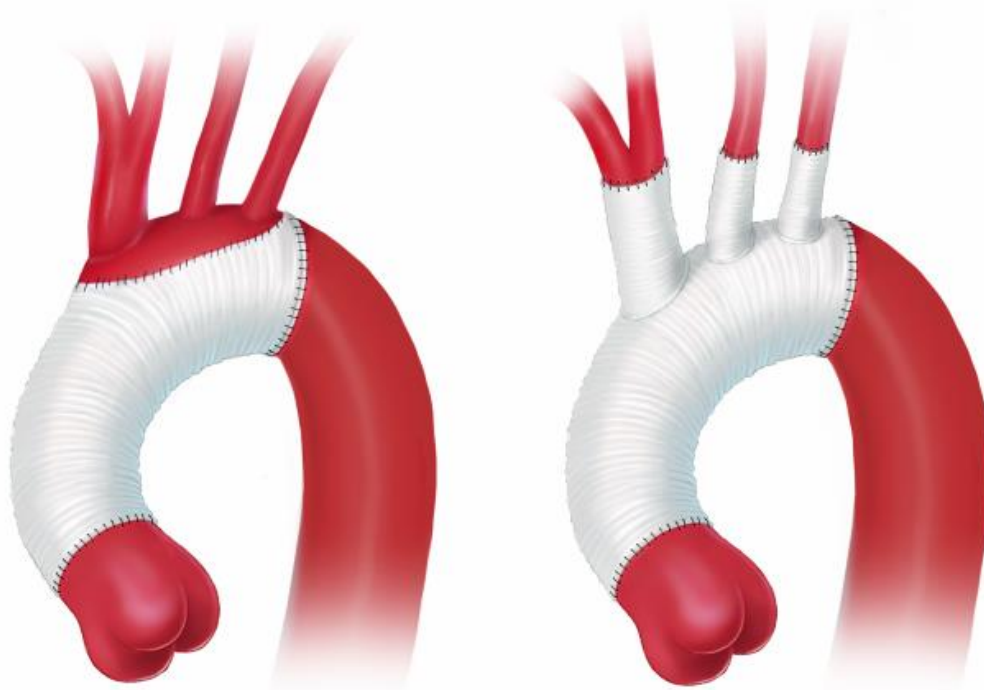
Figure 3



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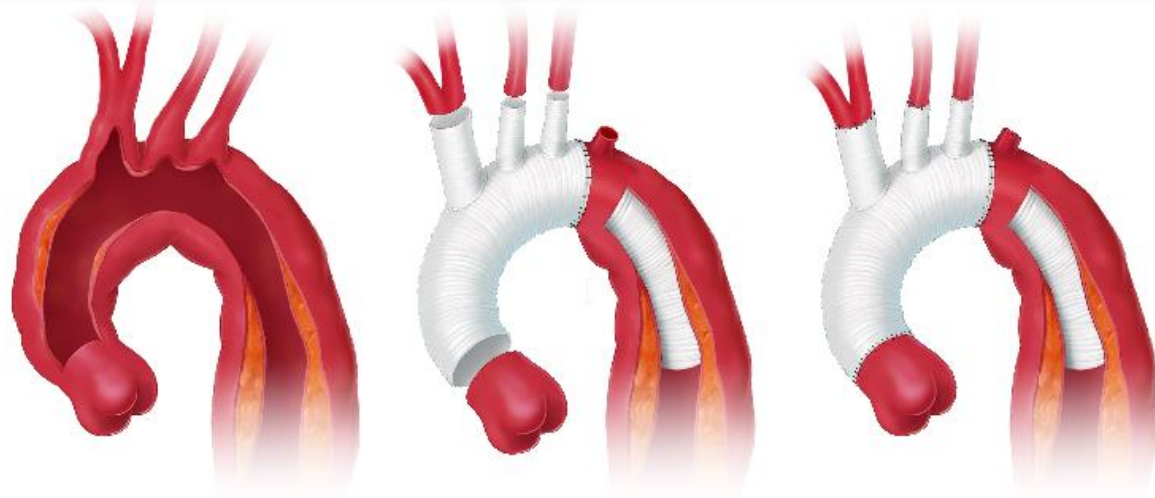
**Figure 4**



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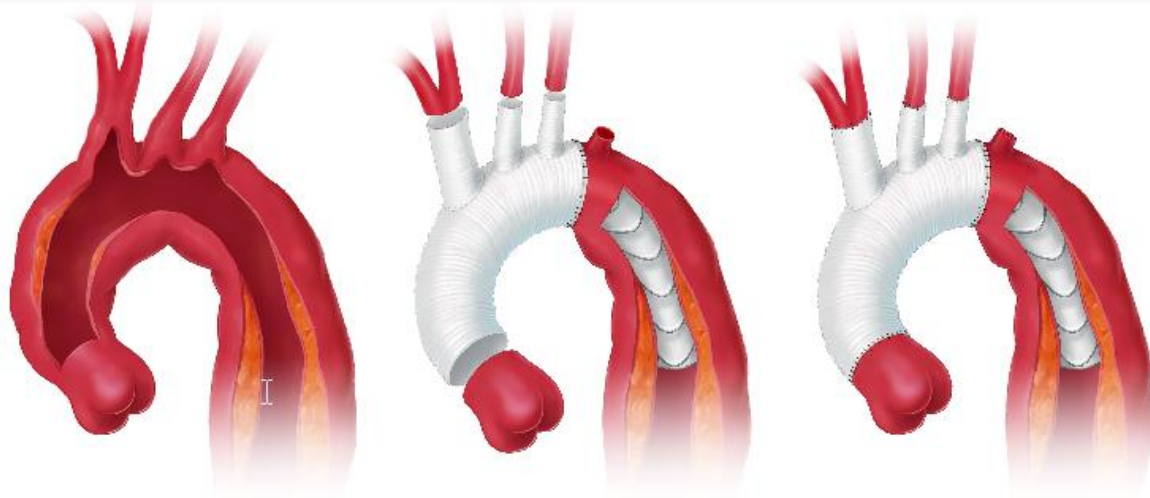
**Figure 5**



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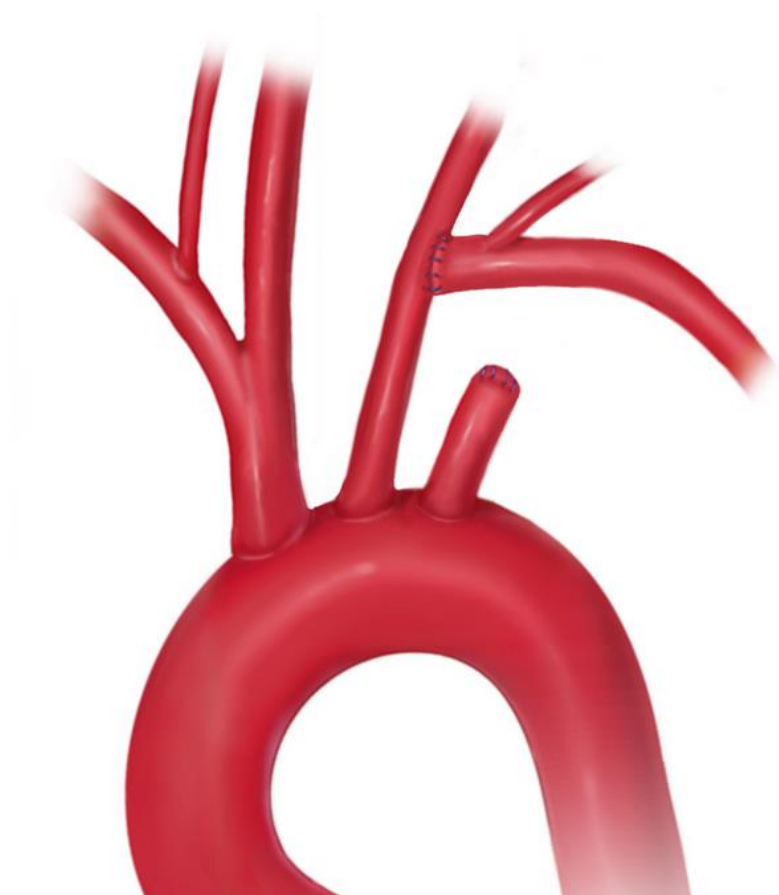
2701  
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Figure 6



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2705  
2706 **Figure 7**



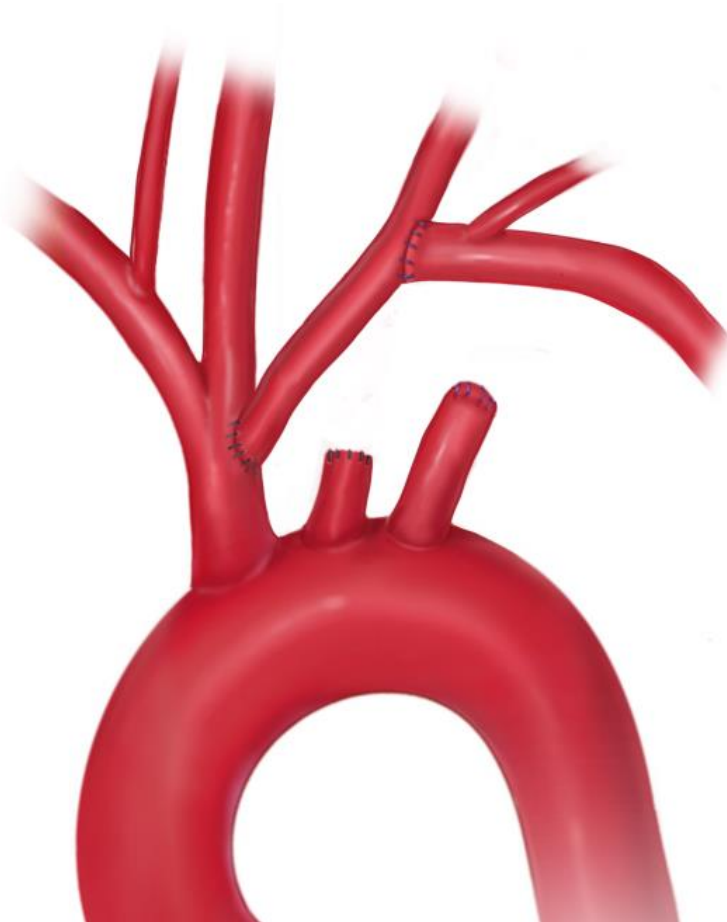
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2710 **Figure 8**



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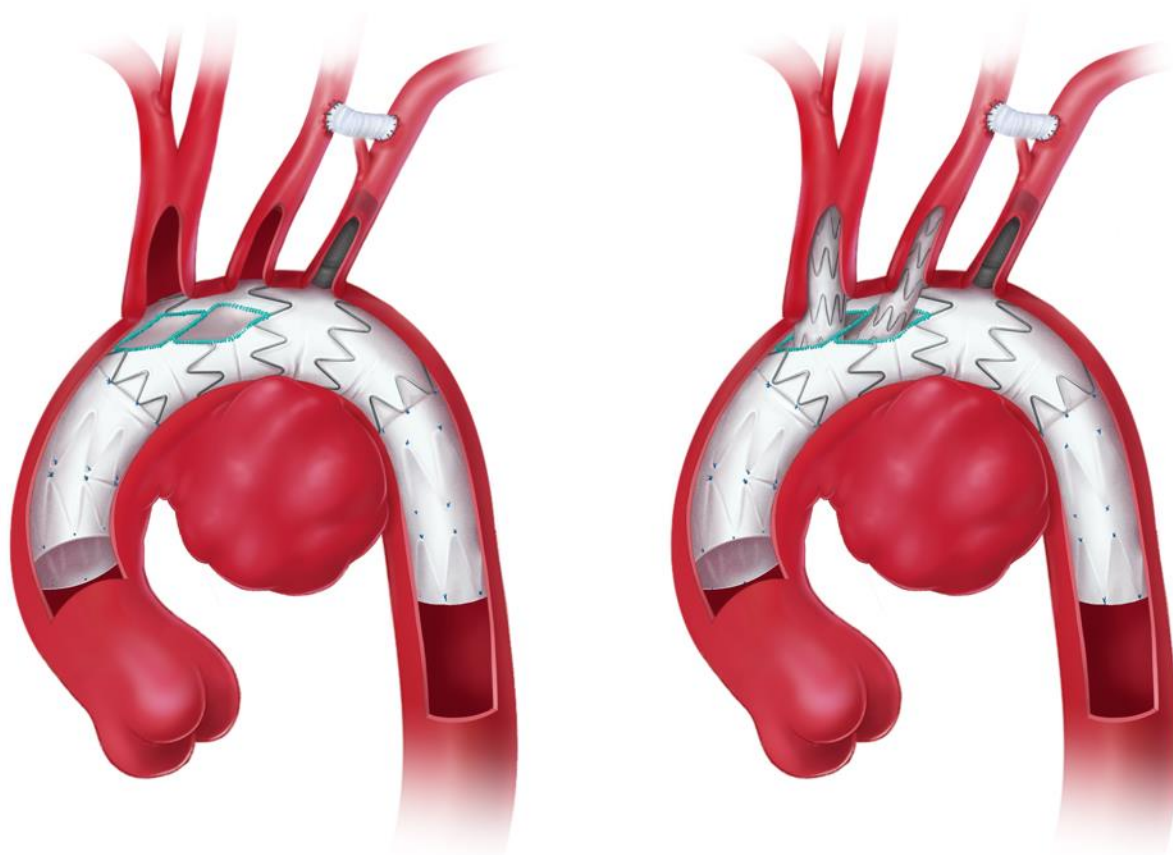
2713  
2714 **Figure 9**



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2717  
2718 **Figure 10**



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